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THE SPINE

A Radiological Text and Atlas

By

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Second Edition, Thoroughly Revised
893 Illustrations on 388 Figures

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Second Edition

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TO
MY WIFE

Preface to the Second Edition

A SECOND edition presents the author with the privilege of a "second look" and the opportunity of bringing his work as close to current concepts as permitted by his efforts and the time which elapses between the last tap on his typewriter and the finished book. Much material, in some measures new and in others a recapitulation and elaboration of already known information, has appeared in the radiological literature since 1955. I have tried to reflect advances in the text and illustrations, and to inspect such information in the light of my own daily experience. It was often difficult to decide just what to include and what to leave out in the selection of new illustrations and the recording of personal observations. Nor could I do justice to the many excellent papers which have appeared, so that of necessity many are not mentioned by name, but are acknowledged with thanks nevertheless.

More than fifty new illustrations have been added, and a number of the original ones have been replaced. The sections on the embryology and radiological anatomy of the spine have been augmented, as has the material on developmental anomalies and structural changes. The myelographic contributions to the diagnosis of discal, intraspinal and vertebral diseases has been reemphasized, and the sections on myelography have been rewritten and, to a great extent, reillustrated. In general, as each chapter was reviewed and revised, I tried to emphasize clinical correlations in accord with the concept of regarding the spine as an organ which reflects systemic disease as well as one having its own peculiar maladies.

The fundamental importance of technical elegance and intelligent planning as indispensable requirements for reliable roentgenological diagnosis is reaffirmed. The current furor over radiation hazards must not be overlooked, and every effort should be made to keep exposure to a minimum. Notwithstanding, economy must never be carried to the point of endangering a patient because of insufficient data or unreliable work. The basic importance of the radiologist as a consultant can be maintained only so long as he actually is one, and he must be personally responsible for work done under his supervision as well as for his own. And insofar as it is humanly possible, that supervision and work should be on the spot and personal, reflecting knowledge of the patient and his complaints as well as the professional approaches to be taken towards a trustworthy opinion. It is a truism that radiological examinations ought to be conducted with the same devoted regard for the patient's welfare as is any other medical discipline. I must confess, however, that sometimes I stare with bemused and saddened admiration while watching the superbly smooth, efficient and impersonal assembly-line mechanics of radiology in busy hospitals. I have often wondered what it would be like to see the Department of Radiology operating like the Department of Surgery, for example, with each patient receiving the undivided attention of his physician. It is apparent that the glow of our viewing boxes still cannot match the focussed, brilliant glare of the operating room lights. The difference between an impersonal approach to the interpretation of roentgenograms and examining a patient requires little comment, and each one of us has to search his soul to see just how he is discharging his responsibility.

During the seven years which have passed since the first edition of *The Spine* I have followed the flow of old and recent ideas and observations in the environment of a new, relatively small but very active general hospital. The cooperative efforts of the various services and their chiefs is acknowledged with thanks. I am particularly grateful to my brother, Dr. Joseph A. Epstein of the division of neurological surgery, Dr. Leroy Lavine of the orthopedic division, and Dr. James I. Berkman, the chief pathologist. They have made my work more pleasant and more exciting by their constant interest, their stimulating collaboration and their warm friendship. My chief technician, Mr. Jerome Sloven, continues to fulfill all the requirements of a master of his craft, and has been assiduous and painstaking in his part of our work. The publishers, Lea & Febiger, again have exhibited their gracious consideration and unfailing helpfulness, for which it is my sincere pleasure to thank them.

BERNARD S. EPSTEIN

NEW HYDE PARK, NEW YORK

Preface to the First Edition

THE vertebral column and its contents are subject to many diseases. Some of these are unique to the spine while others are local manifestations of widespread conditions. Much has been published on the radiology of the spine, most of it in specialized journals. It appeared a good idea to gather available information and review our own material in a work which would treat the spinal column as an entity. In this volume are correlated twenty years of observation of the radiological and clinical variations of vertebral column disease. Inasmuch as an understanding of embryologic development is practically essential for comprehending the infant, growing and congenitally malformed spine, this aspect was presented before going on with the normal radiologic picture. In reviewing the variations seen with the same disease, I have tried to include a concept of progression from the early phases to the full blown syndrome.

The details of roentgenographic technic are not included here. However, I would like to stress the fundamental importance of technical excellence. Not only must the films be of superb quality, but proper views must be selected. It should not be left to a technician's judgment alone to decide which views are required. The radiologist must assume the obligation of planning the study as well as the responsibility of diagnosis. Myelography is basic in investigations of the spinal canal, and has been integrated with other technics rather than treated as a separate entity. The application of body section roentgenography to examination of the vertebral column has been emphasized

In reviewing the literature and compiling a bibliography, it soon became apparent that a volume this size could be filled with references alone. The papers referred to specifically illustrate points under discussion. There are many others of equal excellence not mentioned by name. To those whose work I quoted, and to the many others who were not cited but who nevertheless contributed greatly to this effort, I extend my thanks.

The tremendous amount of work done on the radiology of the vertebral column reflects the concern of neurological surgeons and neurologists, orthopedists, internists, pediatricians and others as well as the radiologist. Much of the neurosurgical and neurological material in this volume was seen with Dr. Leo M. Davidoff, to whom I am grateful for much encouragement and stimulation. Dr. Joseph Milgram and Dr. M. T. Koven were of constant help from the orthopedic aspect. To them, and to the medical staff of The Jewish Hospital of Brooklyn, I offer my appreciation. Most of this work was done while I was a member of the Department of Radiology of The Jewish Hospital of Brooklyn. I extend my thanks to that institution for many years of pleasant work. Contributed illustrations are acknowledged with thanks in the respective subheadings.

My brother, Dr. Joseph A. Epstein, read the manuscript and made many helpful contributions. I acknowledge his help with thanks. Dr. Arthur Sawitsky reviewed the ninth chapter and suggested several changes.

The roentgenograms were made for the most part by Mr. Jerome Sloven, who has been a bulwark of technical help. I thank him for many hours of hard, patient and devoted work. The photographs were made by Mr. Marvin Ehlin, to whom I am obliged for constant cooperation.

The publishers have always been most cooperative and helpful. It is a privilege to work with them, and I am indebted to them for their patience and great skill.

BERNARD S. EPSTEIN, M.D.

NEW HYDE PARK, NEW YORK

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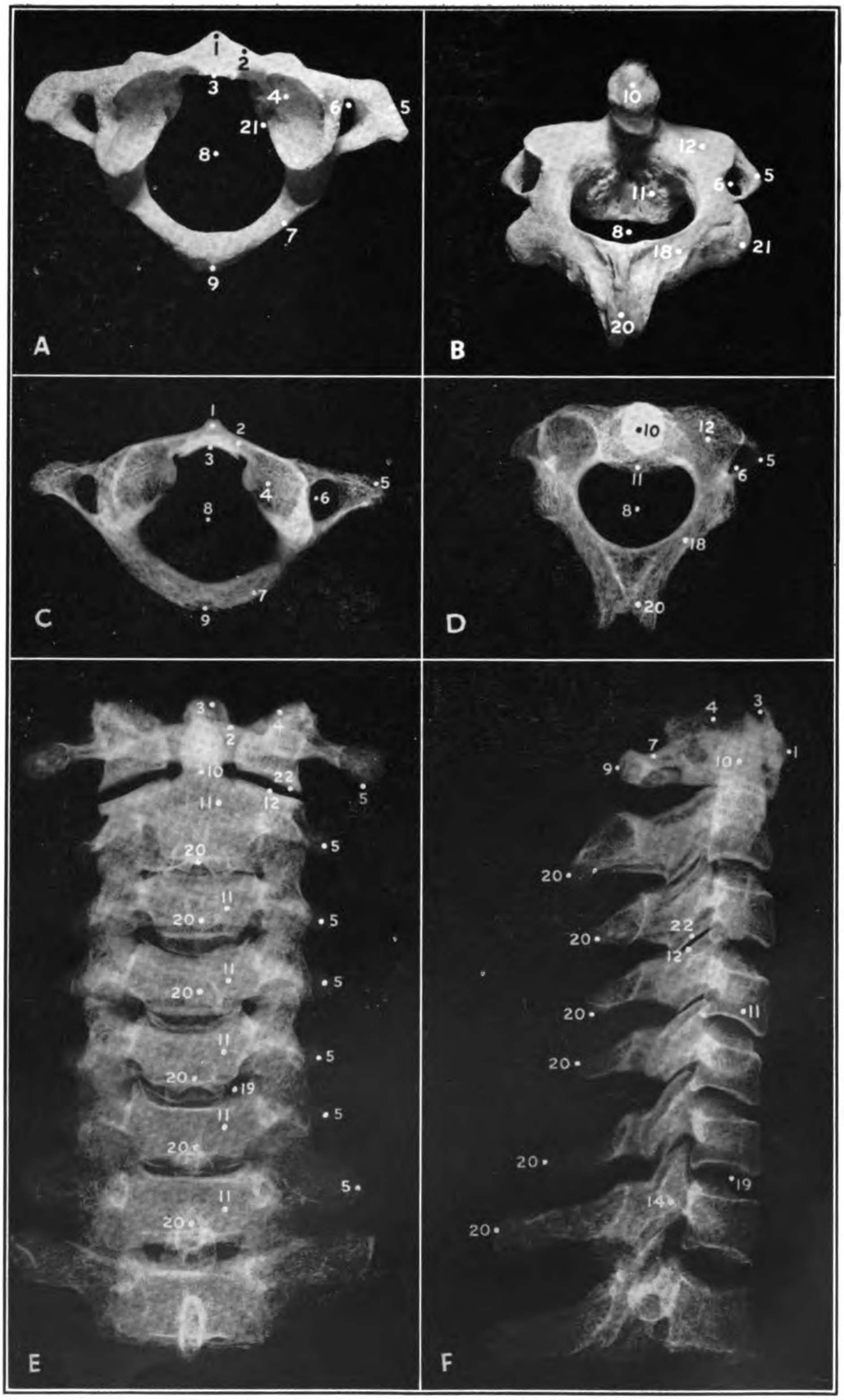


FIG. 1.—Photographs and radiographs of various aspects of dried specimens of the *cervical vertebræ*, that demonstrate their anatomy (1st thoracic vertebra also shows in Fig. 1*E* and *F* and FIG. 2*C*).

A, Photograph of superior aspect of atlas. *B*, Photograph of superior aspect of axis. *C*, Supero-inferior radiograph of atlas. *D*, Supero-inferior radiograph of axis. *E*, Anteroposterior radiograph of articulated vertebræ. *F*, Lateral radiograph of articulated vertebræ.

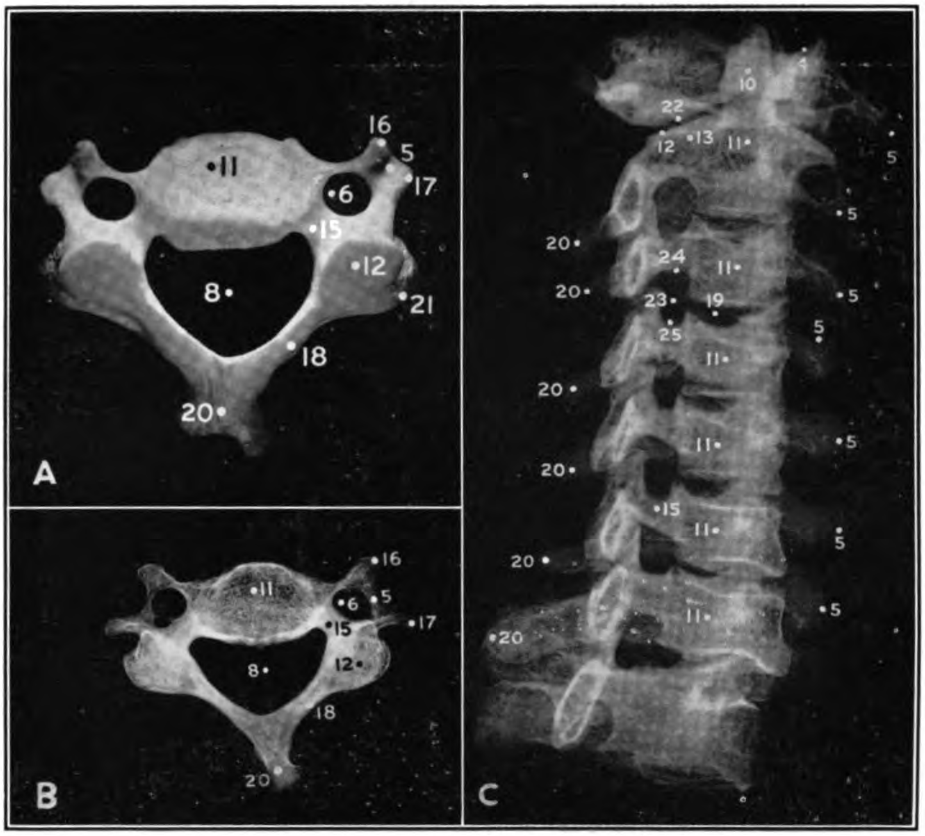


FIG. 2.—*A*, Photograph of superior aspect of fourth cervical vertebra. *B*, Supero-inferior radiograph of fourth cervical vertebra. *C*, Oblique radiograph of of articulated cervical vertebræ.

LEGEND FOR FIGURES 1 AND 2

1. Anterior tubercle of atlas
   (*tuberculum anterius atlantis*)
2. Anterior arch of atlas
   (*arcus anterior atlantis*)
3. Articular facet for odontoid process of axis
   (*fovea dentis*)
4. Superior articular facet of atlas
   (*fovea articularis superior atlantis*)
5. Transverse process
   (*processus transversus*)
6. Transverse foramen
   (*foramen transversarium*)
7. Posterior arch of atlas
   (*arcus posterior atlantis*)
8. Vertebral foramen
   (*foramen vertebrale*)
9. Posterior tubercle of atlas
   (*tuberculum posterius atlantis*)
10. Odontoid process of axis
    (*dens epistropheus*)
11. Body of vertebra
    (*corpus vertebræ*)
12. Superior articular facet
    (*facies articularis superior*)
13. Superior articular process
    (*processus articularis superior*)
14. Lateral mass
    (*massa lateralis*)
15. Pedicle of vertebral arch
    (*radix arcus vertebræ*)
16. Anterior tubercle of cervical vertebræ
    (*tuberculum anterius vertebrarum cervicalium*)
17. Posterior tubercle of cervical vertebræ
    (*tuberculum posterius vertebrarum cervicalium*)
18. Lamina of vertebral arch
    (*lamina arcus vertebræ*)
19. Intervertebral disc space
20. Spinous process
    (*processus spinosus*)
21. Inferior articular process
    (*processus articularis inferior*)
22. Inferior articular facet
    (*facies articularis inferior*)
23. Intervetebral foramen
    (*foramen intervertebrale*)
24. Superior vertebral notch
    (*incisura vertebralis superior*)
25. Inferior vertebral notch
    (*incisura vertebralis inferior*)

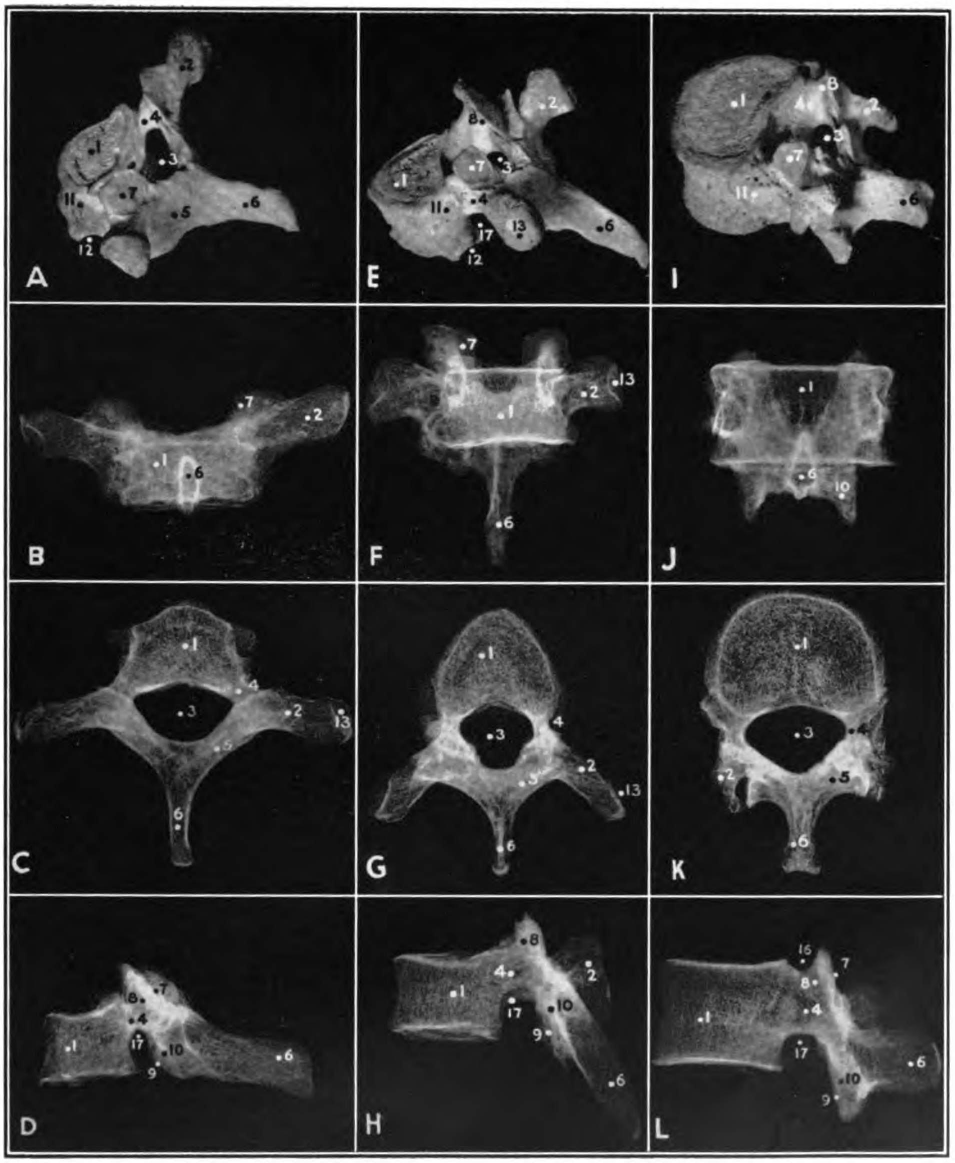


FIG. 3.—Photographs and radiographs of thoracic vertebræ, that illustrate their anatomical details.

1st thoracic vertebra. *A*, photograph; *B*, anteroposterior radiograph; *C*, supero-inferior radiograph; *D*, lateral radiograph.

5th thoracic vertebra. *E*, photograph; *F*, anteroposterior radiograph; *G*, supero-inferior radiograph; *H*, lateral radiograph.

12th thoracic vertebra. *I*, photograph; *J*, anteroposterior radiograph; *K*, supero-inferior radiograph; *L*, lateral radiograph.

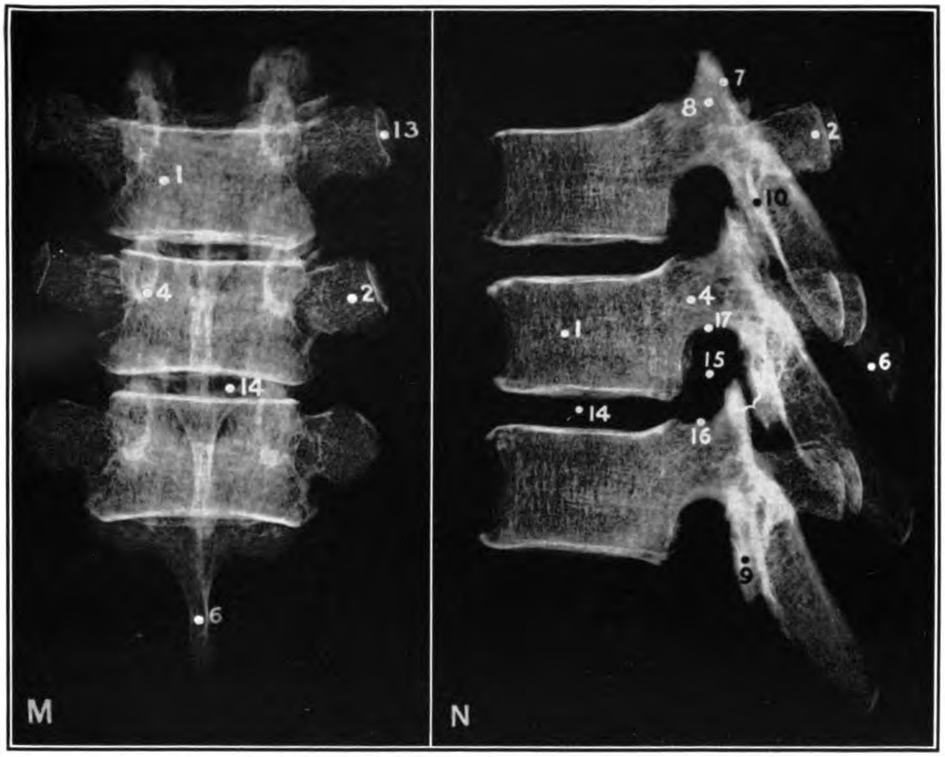


FIG. 3. (*Continued*)—6th, 7th, and 8th thoracic vertebræ, articulated. *M*, anteroposterior radiograph; *N*, lateral radiograph.

LEGEND FOR FIGURE 3

1. Body of vertebra
   (*corpus vertebræ*)
2. Transverse process
   (*processus transversus*)
3. Vertebral foramen
   (*foramen vertebrale*)
4. Pedicle of vertebral arch
   (*radix arcus vertebræ*)
5. Lamina of vertebral arch
   (*lamina arcus vertebræ*)
6. Spinous process
   (*processus spinosus*)
7. Superior articular facet
   (*facies articularis superior*)
8. Superior articular process
   (*processus articularis superior*)
9. Inferior articular facet
   (*facies articularis inferior*)
10. Inferior articular process
    (*processus articularis inferior*)
11. Superior costal facet
    (*fovea costalis superior*)
12. Inferior costal facet
    (*fovea costalis inferior*)
13. Costal facet for tubercle of rib
    (*fovea costalis transversalis*)
14. Intervertebral disc space
15. Intervertebral foramen
    (*foramen intervertebrale*)
16. Superior vertebral notch
    (*incisura vertebralis superior*)
17. Inferior vertebral notch
    (*incisura vertebralis inferior*)
18. Intervertebral articulation
    (*Bracket is used in illustration to indicate articulation*)

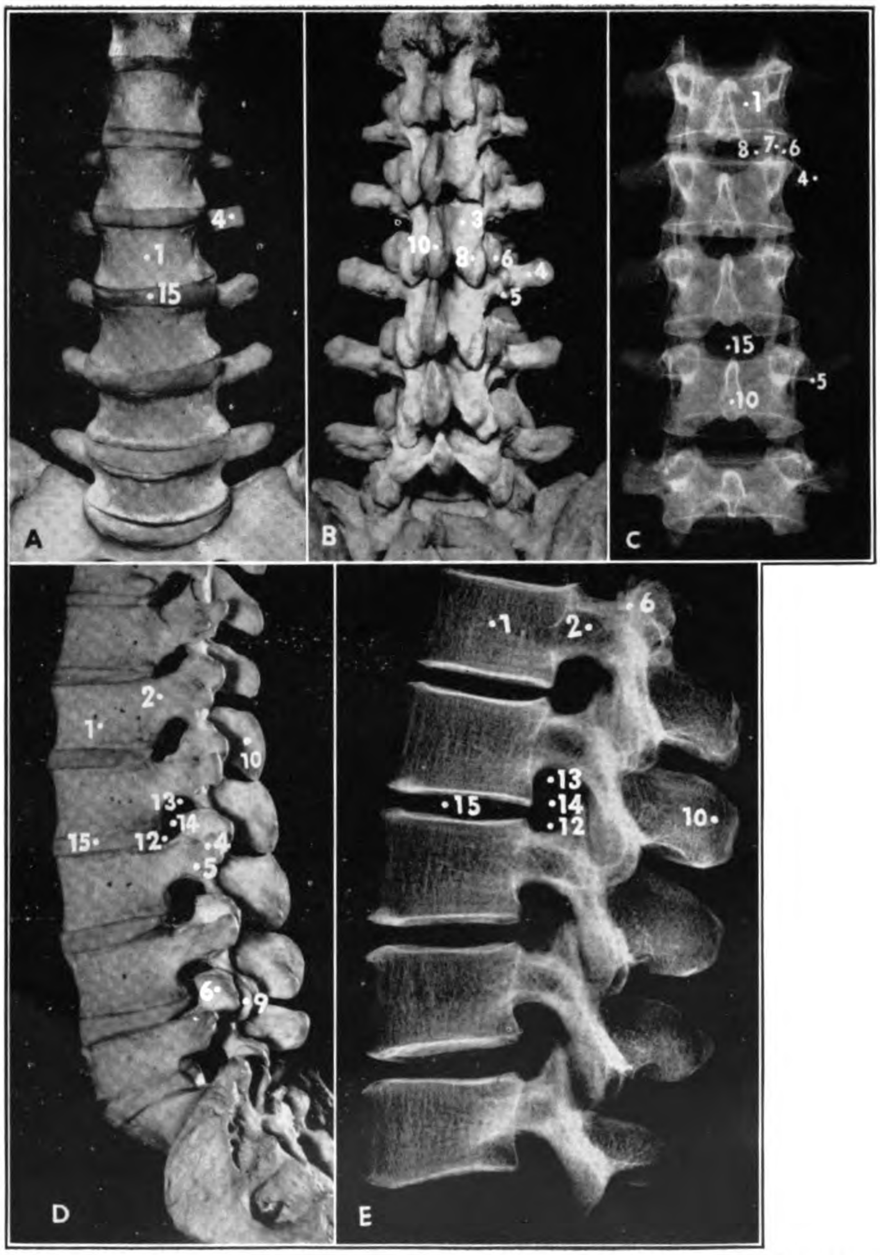


FIG. 4.—Photographs and radiographs of dried speciments of articulated lumbar vertebræ, that illustrate their anatomic details.

A, Photograph of anterior aspect; *B*, photograph of posterior aspect; *C*, anteroposterior radiograph; *D*, photograph of lateral aspect; *E*, lateral radiograph.

LEGEND FOR FIGURES 4 AND 5

1. Body of vertebra (*corpus vertebræ*)
2. Root (pedicle) of vertebral arch (*radix arcus vertebræ*)
3. Lamina of vertebral arch (*lamina arcus vertebræ*)
4. Transverse process (*processus transversus*)
5. Accessory process (*processus accessorius*)
6. Superior articular process (*processus articularis superior*)
7. Superior articular facet (*facies articularis superior*)
8. Inferior articular process (*processus articularis inferior*)
9. Inferior articular facet (*facies articularis inferior*)
10. Spinous process (*processus spinosus*)
11. Vertebral foramen (*foramen vertebrale*)
12. Superior vertebral notch (*incisura vertebralis superior*)
13. Inferior vertebral notch (*incisura vertebralis inferior*)
14. Intervertebral foramen (*foramen intervertebrale*)
15. Intervertebral disc space

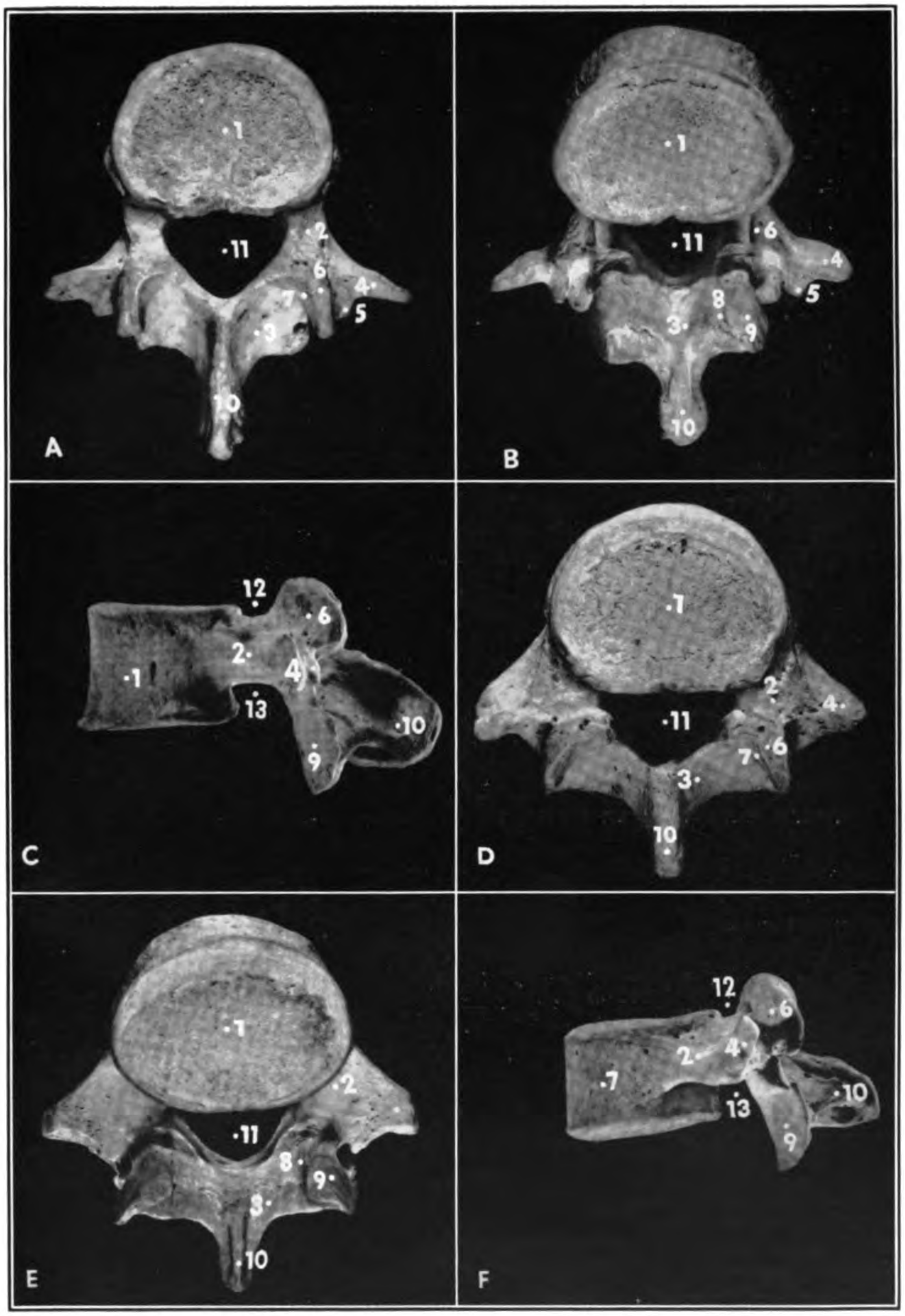


FIG. 5.—Photographs of dried specimens of lumbar vertebræ, that illustrate their anatomic details.

3d lumbar vertebra: *A*, superior aspect; *B*, inferior aspect; *C*, lateral aspect.

5th lumbar vertebra: *D*, superior aspect; *E*, inferior aspect; *F*, lateral aspect.

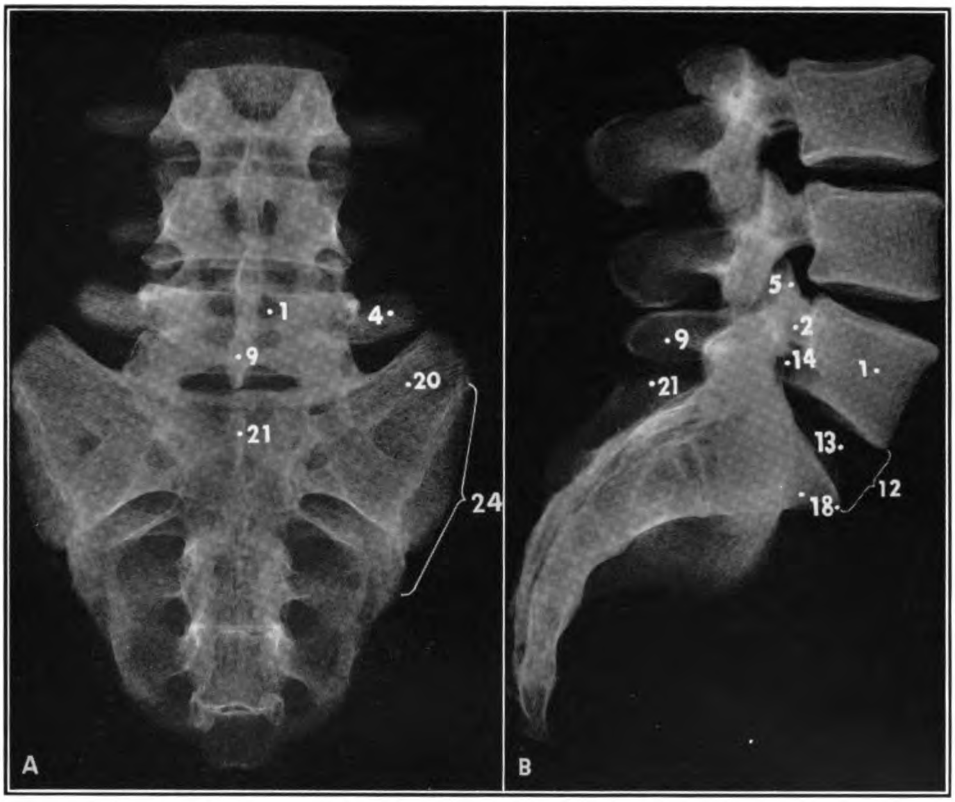


FIG. 6.—Radiographs of articulated lumbar vertebræ and sacrum. *A*, anteroposterior radiograph; *B*, lateral radiograph.

LEGEND FOR FIGURES 6 AND 7

5TH LUMBAR VERTEBRA

1. Body of vertebra
   (*corpus vertebræ*)
2. Root of vertebral arch
   (*radix arcus vertebræ*)
3. Lamina of vertebral arch
   (*lamina arcus vertebræ*)
4. Transverse process
   (*processus transversus*)
5. Superior articular process
   (*processus articularis superior*)
6. Superior articular facet
   (*facies articularis superior*)
7. Inferior articular process
   (*processus articularis inferior*)
8. Inferior articular facet
   (*facies articularis inferior*)
9. Spinous process
   (*processus spinosus*)
10. Superior vertebral notch
    (*incisura vertebralis superior*)
11. Inferior vertebral notch
    (*incisura vertebralis inferior*)

LUMBOSACRAL JUNCTION

12. Articulation for the vertebral bodies
    (Bracket is used in illustration
    to indicate articulation)
13. Intervertebral disc
    (*space*)
14. Intervertebral foramen
    (*foramen intervertebrale*)
15. Articulation for the vertebral arches
    (Bracket is used in illustration
    to indicate articulation)

1ST SACRAL VERTEBRA

16. Base
    (*basis ossis sacri*)
17. Body of vertebra
    (*corpus vertebræ*)
18. Promontory
    (*promontorium*)
19. Sacral canal
    (*canalis sacralis*)
20. Lateral mass
    (*pars lateralis*)
21. Spinous process
    (*processus spinosus*)
22. Superior articular process
    (*processus articularis superior*)
23. Superior articular facet
    (*facies articularis superior*)
24. Iliac articular surface
    (*facies auricularis*)
25. Anterior sacral foramen
    (*foramen sacralis anterior*)
26. Posterior sacral foramen
    (*foramen sacralis posterior*)

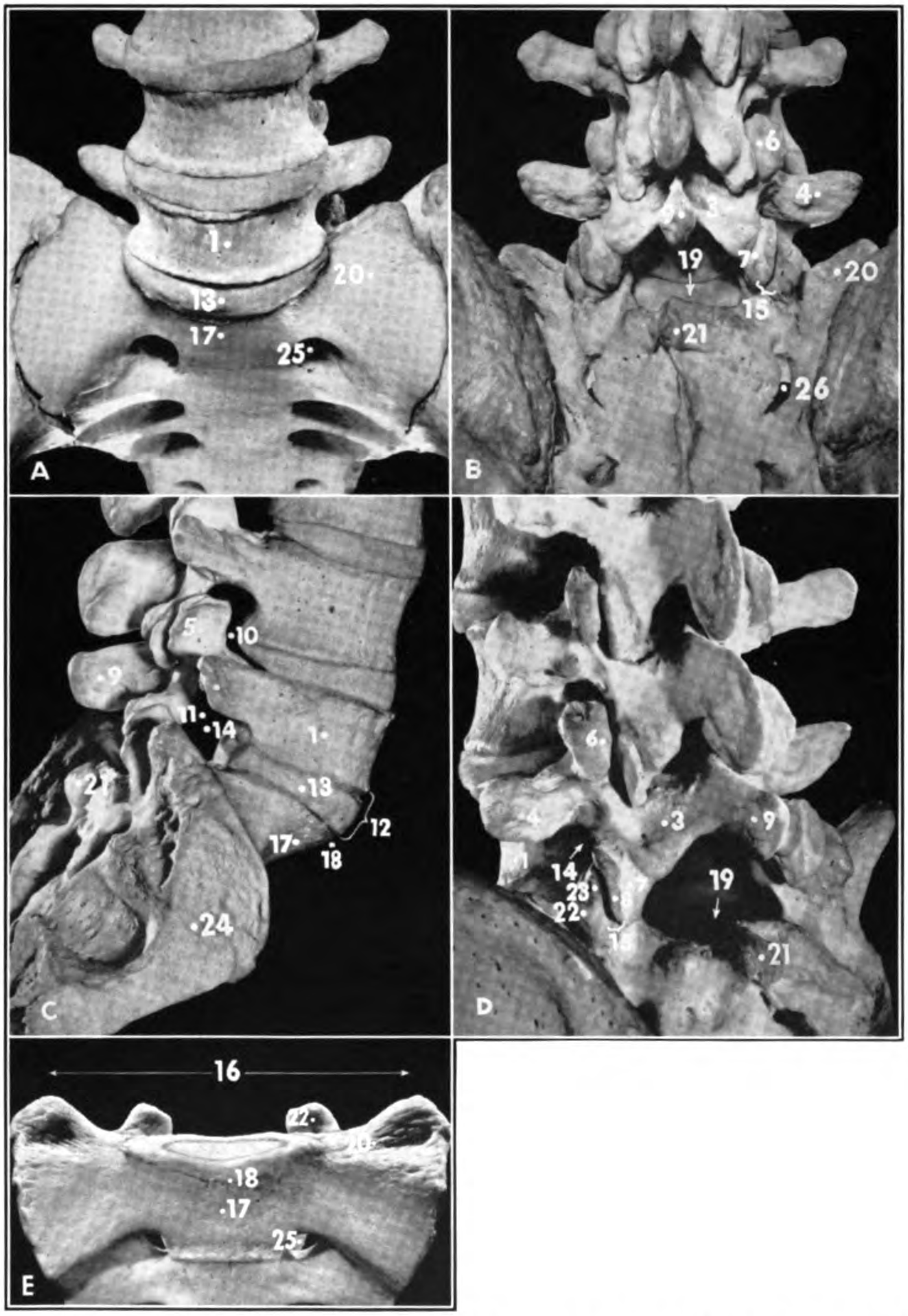


FIG. 7.—Photographs and radiographs of dried specimens of articulated lumbar vertebræ and sacrum, illustrating the anatomy of these structures and the lumbosacral junction.

A, photograph of anterior aspect; *B*, photograph of posterior aspect; *C*, photograph of lateral aspect; *D*, photograph of lateroposterior aspect; *E*, photograph of anterior aspect of superior portion of sacrum.

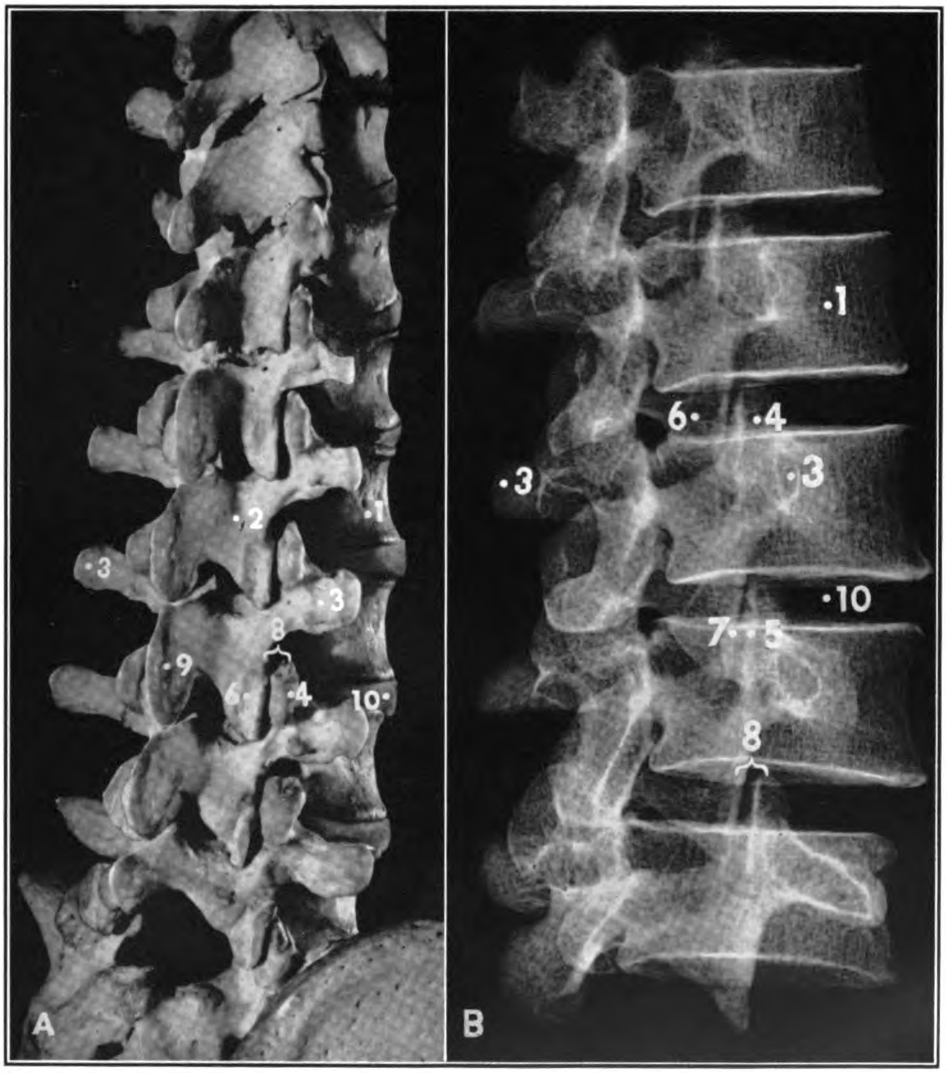


FIG. 8.—Photograph and radiograph of dried specimens of articulated lumbar vertebræ: *A*, photograph of lateroposterior aspect; *B*, lateroposterior radiograph.

LEGEND FOR FIGURE 8

1. Body of vertebræ
   (*corpus vertebræ*)
2. Lamina of vertebral arch
   (*lamina arcus vertebræ*)
3. Transverse process
   (*processus transversus*)
4. Superior articular process
   (*processus articularis superior*)
5. Superior articular facet
   (*facies articularis superior*)
6. Inferior articular process
   (*processus articularis inferior*)
7. Inferior articular facet
   (*facies articularis inferior*)
8. Intervertebral articulation
   (*Bracket is used in illustration to indicate articulation*)
9. Spinous process
   (*processus spinosus*)
10. Intervertebral disc space

---

* FIGS. 1 to 8.—From Radiography and Clinical Photography, William S. Cornwell, Editor; with permission of Eastman Kodak Company.

Chapter

1

Normal Spine

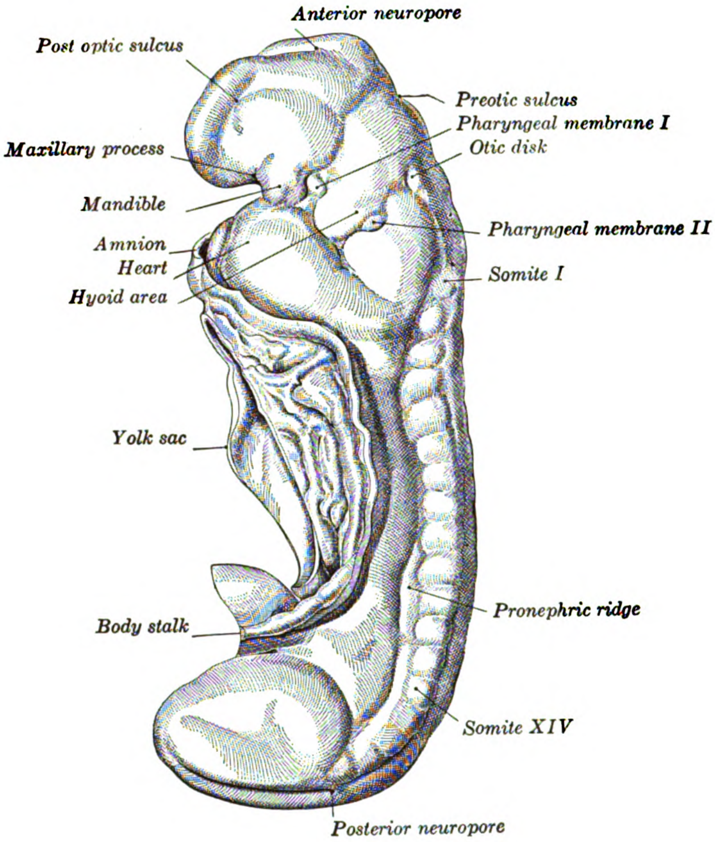
EMBRYOLOGIC CONSIDERATIONS

FOLLOWING fertilization of the ovum, the male and female pronuclei combine into the new segmentation nucleus, which further divides into the two cell stage, beginning the growth of a new individual. Passing through the morula stage, an outer layer of cells, the trophoblast, and an inner cluster, the inner cell mass, evolve. Secretion of fluid by the trophoblast into the morula results in the blastocyst, a vesicle formed by the trophoblast to which the inner cell mass is attached. At about the eighth day after fertilization the blastocyst becomes imbedded in the endometrium, and is completely covered over by the twelfth day. The trophoblast combines with the mesoderm, forming the chorion, the protecting coat which relates the fetus to the maternal nutritional and excretory resources.

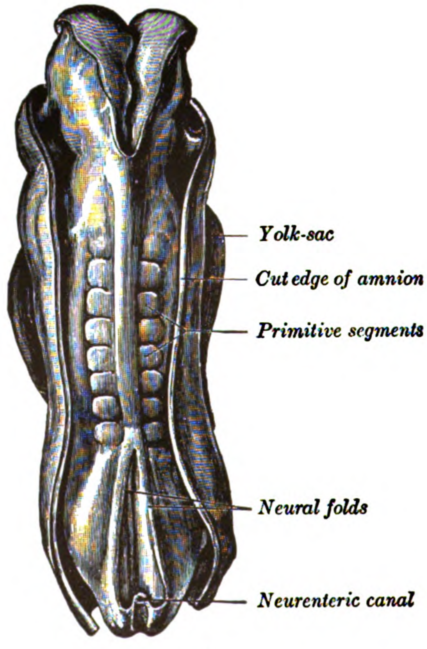
During this time the inner cell mass enlarges, developing into the outer ectodermal and inner entodermal layers. These rapidly separate, the ectoderm forming the amnion and its contained amniotic cavity, while the entoderm constitutes the yolk sac protruding into the blastocyst. The embryo itself develops from the germ disc, that portion of the ectoderm and entoderm which remain in apposition. The extraembryonal mesoderma layer evolving from cells migrating from the borders of this disc form a loose network of cellular processes stretching across the blastocyst cavity and occupy the space between the trophoblast and yolk sac. The entire embryo then becomes suspended in the chorionic cavity by the mesodermal primitive body stalk.

Further growth of the embryo procedes from the embryonic disc. A primitive groove appears in the midline, beneath which the rapidly growing primitive streak develops. These cells later form the definitive mesoderm, part of which migrates to the cephalic end of the embryo, where it comes in contact with the primitive node of Hensen, another cluster of mesodermal cells. The notochord develops from Hensen's node as a medially situated rod. At the end of two weeks the embryo is about 1.5 mm. long, somewhat oval in shape, and the three germ layers are well differentiated. Cephalic to Hensen's node thickened, rounded edges of ectoderm appear as the neural folds on either side of the notochord and prochordal plate, evolving into the neural groove. Further growth converts the groove into a tube. The cephalic end, which develops more rapidly, expands to form the brain, while the caudal portion develops as the spinal cord.

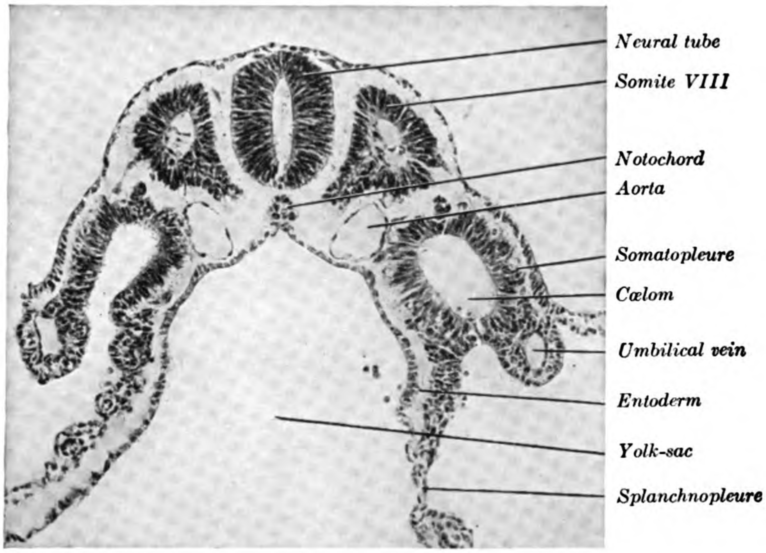
By the third week the cephalic end of the embryo is marked by the prominence of the neural plate. Thereafter the paraxial bar, the mesodermal column on either side of the notochord becomes segmented into triangular-shaped symmetrically disposed somites, situated so that their bases are medial and apices lateral in position. From these the skeletal, muscular


Anterior neuropore
Post optic sulcus
Preotic sulcus
Pharyngeal membrane I
Otic disk
Maxillary process
Mandible
Pharyngeal membrane II
Amnion
Heart
Hyoid area
Somite I
Yolk sac
Pronephric ridge
Body stalk
Somite XIV
Posterior neuropore

A


Yolk-sac
Cut edge of amnion
Primitive segments
Neural folds
Neurenteric canal

B



C

FIG. 9.—*A*, Lateral view of a 14-somite human embryo. × 50. *B*, Dorsum of human embryo. 2.11 mm. in length. *C*, Transverse section through somite VIII (primitive segments of 14-somite human embryo. (Gray's *Anatomy*, Lea & Febiger.)

and nervous systems arise. The somites appear first at the cephalic end and progress caudally. Each has a cavity called the myocele, which soon fills with proliferating cells. The coelomic cavity appears lateral to these, with somatopleure dorsally and splanchopleure ventrally. At the junction of the lateral mesoderm and somites a strand of cells, termed the intermediate cell mass, later forms the genitourinary system.

The vertebral column evolves around the notochord. Lying on the ventral aspect of the neural tube, it extends from the caudal extremity to the midbrain. In the 15 mm. fetus its cephalic end passes through the basisphenoid to the pharyngeal surface, and reenters the sphenoid to terminate at the dorsum sellæ. The primitive vertebral segments are separated from each other by intersegmental septæ. Each segment differentiates into three groups of cells. The cutis-plate, or dermatome, forms on the dorsolateral aspect of the myocele, and forms the skin. The muscle-plate, or myotome, is situated on the medial side of the myocele, forms the muscles of the segment. From the core of the myocele, next to the notochord, arises the sclerotome, the sclaerotogenous layer of cells which rapidly proliferate to surround the notochord and neural tube with a continuous mesodermal sheath termed the membranous vertebral column. The sclerotome divides into cranial and caudal parts by a transverse fissure, the sclerotomic fissure, which is transient and soon vanishes. The original somite can be identified in these two portions, the anterior aspect formed by loosely arranged cells while the posterior half is more compact. Between these the intervertebral disc is formed, in the vicinity of the sclerotomic fissure. Cells from the denser posterior mass grow into the spaces between the myotomes of the adjacent and caudal segments, extending dorsally and ventrally. The dorsal segments form the neural arches, while the ventral segments extend into the body wall as the transverse processes. The vertebral bodies are formed by fusion of the caudally placed dense segment of one sclerotome and the anterior portion of the next caudal sclerotome. The vertebræ alternate in position with the myotomes. Muscles arising from one myotome is related thus to two skeletal elements and the interposed intervertebral disc. The

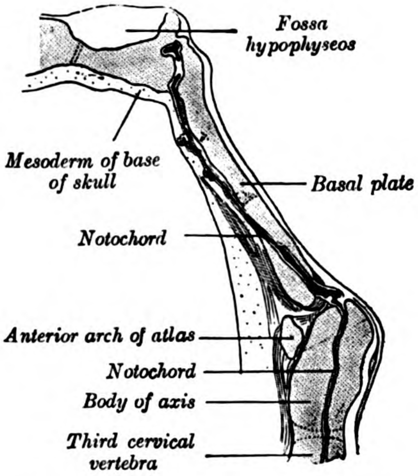


FIG. 10.—Sagittal section of cephalic end of notochord. (Gray's *Anatomy*, Lea & Febiger.

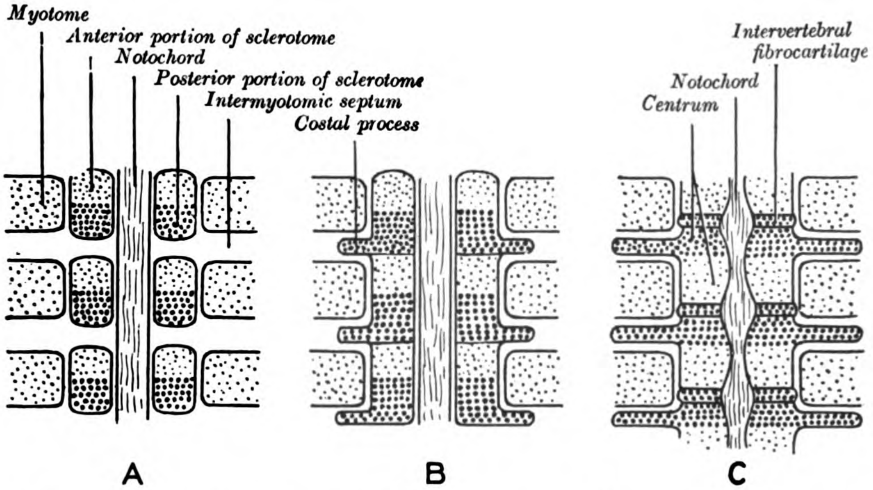

Myotome
Anterior portion of sclerotome
Notochord
Posterior portion of sclerotome
Intermyotomic septum
Costal process
Intervertebral fibrocartilage
Notochord
Centrum
A
B
C

FIG. 11.—Scheme showing the manner in which each vertebral centrum is developed from portions of two adjacent segments. (Gray's *Anatomy*, Lea & Febiger.)

annulus fibrosus forms from the peripheral portions of the primitive discs. The intersegmental arteries, which arise from the primitive aorta, appear very early just caudal to the dense zone of mesenchymal cells, and finally are placed in the midportion of the vertebral bodies.

This so-called "membranous vertebral column" is succeeded at about the fourth week of fetal life by the cartilaginous vertebral column. Two cartilaginous centers separated by the ventrodorsal extension of the perichordal sheath appear on either side of the notochord, and rapidly extend around it, thereby forming the bodies of the cartilaginous vertebræ. The perichordal sheath quickly disappears at this stage. Second pairs of cartilaginous foci appear in the lateral parts of the vertebral arch, grow backward on either side of the neural tube to form the cartilaginous vertebral arch, and still separate cartilaginous centers appear for each costal arch.

The notochord, which remains unchanged as a solid cord of uniform size during the mesenchymal period, is progressively compressed into the regions of the dense intervertebral discs. This is believed due to the increased pressure prevailing during the rapid enlargement of the cartilaginous bodies.

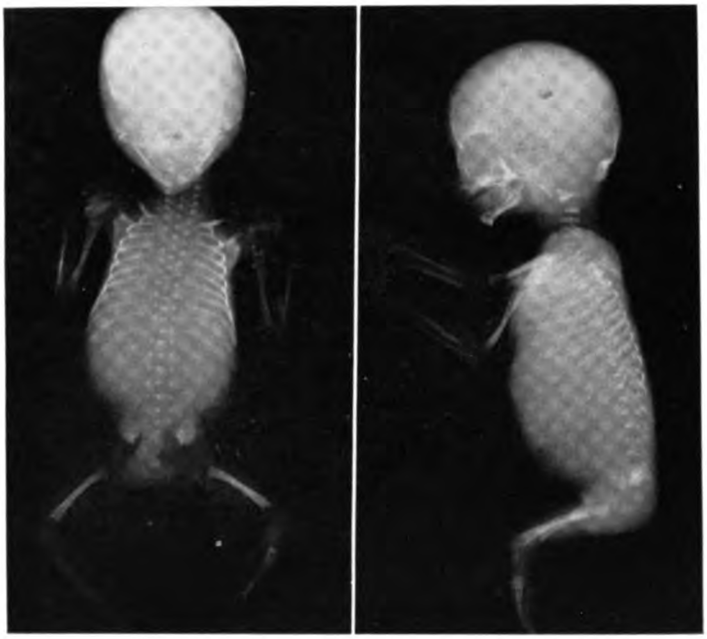


FIG. 12.—Anteroposterior and lateral views of three month fetus. Three main ossification centers are seen in the anteroposterior view, and a single center is visible in the vertebral bodies in the lateral view.

The dense mesenchymal anlage of the annulus fibrosus becomes markedly reduced during chondrification of the bodies, but the outermost rim of the disc dividing the cartilaginous bodies persists.

By the seventh to eighth week of gestation, the cartilaginous cells in the centers of the vertebral bodies become surrounded by an interstitial matrix. During this stage the anterior and posterior longitudinal spinal ligaments develop. The anterior longitudinal ligament is firmly attached to the cartilaginous vertebral bodies, while the posterior longitudinal ligament is attached to the intervertebral disc tissue, but is not as closely attached to the posterior surfaces of the vertebræ.

As chondrification proceeds, the notochordal cells are progressively squeezed out from the vertebral bodies and displaced into the intervertebral discs. Here they become enclosed by a dense rim of cells which have not undergone chondrification and which form the true annulus fibrosus. During this period, some mucoid degeneration and proliferation occurs in the notochordal cells, which then forms the nucleus pulposus. During this migration of notochordal tissue the annulus fibrosus becomes larger. The perichordal sheath itself remains in the center of the cartilaginous vertebræ and is represented by a mucoid streak.

At the ninth week of fetal life anterior and posterior indentations into the cartilaginous body are produced by periosteal vessels. Soon after this the cartilage is invaded by these vessels, which produce ventral and dorsally situated blood lakes. Early in the embryologic development of the vertebral body, ossification centers are formed dorsally and ventrally in the vertebral body, separated by cartilaginous septæ which soon disappear. Ossification proceeds rapidly and is well seen by the time the embryo reaches four months of age.

The early centers of ossification of the vertebral bodies lie dorsally and ventrally. They do not correspond with the centers of chondrification of the cartilaginous stage, the latter being situated to the left and right of the ventro-dorsal extension of the perichordal sheath. The anterior and posterior centers fuse early and form one large center of ossification for the vertebral body. The first centers appear in the lower thoracic and upper lumbar regions and rapidly extend cranially, less rapidly caudally. As the dorsal blood lakes and the center of ossification enlarge, it displaces the mucoid streak ventrally, eventually destroying this structure. Occasionally some remnants of the mucoid streak may remain in the vertebral body. The then-formed centrally situated body nucleus gives off capillary vessels which resorb the surrounding cartilage in a stellate manner.

At approximately the fifth to sixth month of fetal life, the ossification center has divided the cartilaginous body into two thick cartilaginous plates which show endochondral ossification towards the intervertebral disc side. Anteriorly and posteriorly, large osseous channels form the entrance of the vertebral vessels. Along the anterior and lateral periphery of the vertebral bodies, horseshoe-shaped cartilaginous plates appear which represent the cartilaginous ring apophyses. Later these form the anlage of the bony ring apophyses which appear in adolescent life. This cartilaginous ring is the seat of some of the fibers of the anterior and lateral aspects of rhe annulus fibrosus, which later are incorporated as Sharpey's fibers at the time of ossification. The posterior margin of the cartilaginous plate and the annulus fibrosus is not involved in this arrangement.

Ossification of the bodies of the vertebræ does not extend to the bony structures of the entire vertebral body. The postero-lateral portions of the vertebral body are ossified by extensions from the vertebral arch centers. During the first few years of life, therefore, the vertebral bodies show two synchondroses termed "neurocentral synchondroses."

Ossification of the vertebral arches begins at approximately the eighth week of fetal life, appearing first in the upper cervical vertebræ, and gradually extending down the spinal column. Each lateral half of the vertebral arch originates from a separate center of ossification, and the vertebral body originates from a third center of ossification.

The rami of the lumbar neural arches unite during the first year of life, followed by similar changes in the neural arches of the thoracic and cervical regions. The neurocentral synchondroses of the cervical vertebræ are joined to the arches on either side at about the third year of life, and similar union in the lower lumbar vertebræ is not complete until about the sixth

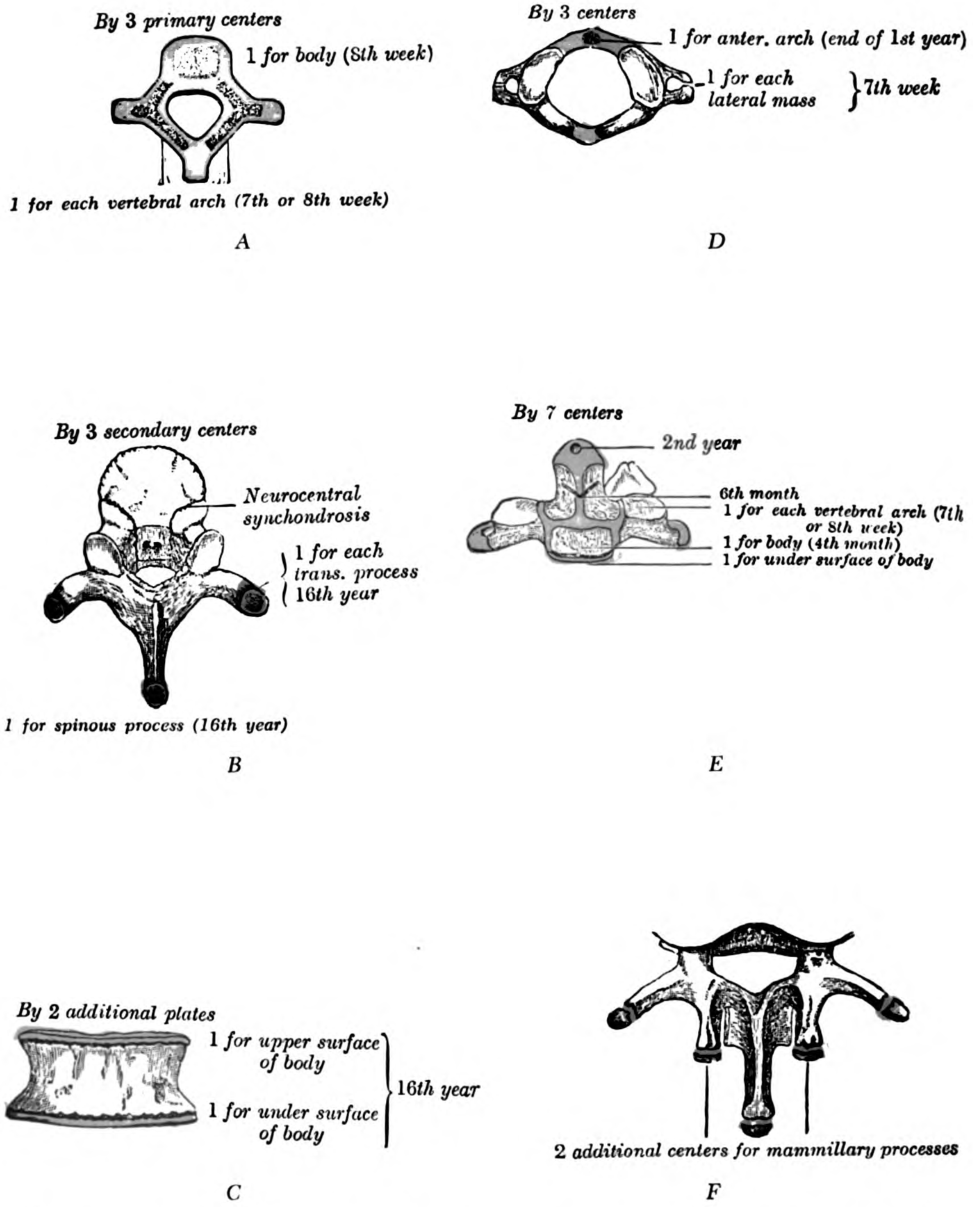

By 3 primary centers
1 for body (8th week)
1 for each vertebral arch (7th or 8th week)
A
By 3 centers
1 for anter. arch (end of 1st year)
1 for each lateral mass
7th week
D
By 3 secondary centers
Neurocentral synchondrosis
1 for each trans. process
16th year
1 for spinous process (16th year)
B
By 7 centers
2nd year
6th month
1 for each vertebral arch (7th or 8th week)
1 for body (4th month)
1 for under surface of body
E
By 2 additional plates
1 for upper surface of body
1 for under surface of body
16th year
C
2 additional centers for mammillary processes
F

FIG. 13.—*A*, Ossification of vertebra at 8 weeks. Three primary centers. *B*, Secondary centers at transverse and spinous processes at sixteenth year. *C*, Apophyseal plates at upper and under surfaces. *D*, Ossification of atlas. *E*, Ossification of axis. *F*, Two additional centers for mamillary processes. (Gray's *Anatomy*, Lea & Febiger.)

year. The tips of the transverse spinal processes remain cartilaginous in the years before puberty. At about the sixteenth year secondary centers appear at the tips of the transverse processes, the tips of the spinous processes, and at the upper and lower surfaces of the vertebral body.

The development of the axis and atlas is rather different from the other cervical segments. The atlas usually ossifies from three centers, one for each lateral mass, which appear at about the seventh week of fetal life, and one for the anterior arch which may be seen soon after birth in about 20 per cent of cases on neck roentgenograms. These two lateral bony masses are separated by a cartilaginous bar, in which a third center of ossification appears at about the end of the first year of life in the other 80 per cent (Tompsett and Donaldson, 1941).

The axis, or epistropheus, ossifies from five primary and two secondary centers. The body of the second cervical vertebra and its neural arch ossifies in much the same manner as the other vertebræ, namely from a single center for the body at about the fourth or fifth month and centers for each lateral half of the vertebral arch at about the second month of gestation. The odontoid process is formed by an upthrust cartilaginous projection from the body of the vertebra. At about the sixth month of fetal life two laterally situated centers of ossification appear in the base of this process. At birth these unite into a single column at the apex of which a cleft remains, for which a separate center of ossification appears at about two years of age. A cartilaginous plate separates the base of the odontoid process from the body of the second cervical vertebra. This gradually becomes ossified, and disappears at or somewhat before adolescence, at which time the apical tip of the odontoid also unites. Ossification involves the circumference of the odontoid process, so that cartilaginous material may persist in its center until late in life.

The transverse processes of the seventh cervical vertebræ sometimes possess a secondary center of ossification from which false ribs form if they do not unite with the true transverse process. Separate centers of ossification for the transverse processes of the fourth to sixth cervical vertebræ have also been described.

Two additional centers of ossification are also present in the lumbar vertebræ. These correspond to the mammilary processes, small prominences on the posterolateral aspects of the superior articular processes. The transverse processes of the first lumbar vertebra sometimes develops from separate centers of ossification. If these do not eventually unite with the vertebral body, a so-called lumbar rib may result. This is a rather uncommon anomaly, but may cause some confusion in identification of vertebral segments. This may be resolved by counting vertebræ on suitable films from the atlas down.

Ossification of the sacral vertebræ proceeds from a primary center and two epiphyseal plates, one for the superior and the other for the inferior surface. Each vertebral arch of the sacrum is ossified from two centers. Lateral to the anterior sacral foramens are two additional centers of ossification for each of the upper three sacral vertebræ. Ossification of the central part of the bodies of the upper three sacral vertebræ appears at about the ninth week of fetal life, and between the sixth and eighth month for the fourth and fifth sacral vertebræ. The costal centers for the lateral parts of the sacral vertebræ appear at about the sixth to eighth month of life at about which time ossification of the vertebral arches also appear. During infancy, the sacral vertebræ are separated from each other by intervertebral fibrocartilages. The lowermost two of these unite at about the eighteenth year of life, and union gradually proceeds so that by adult life the entire sacrum is firmly united. The sites of these intervertebral fibrocartilages can be observed on sagittal sections of anatomic specimens of the sacrum. Radiologically, they can be identified on antero-posterior tilt-up studies of the sacrum as horizontally placed, spindle-shaped radiolucent areas, sometimes containing small calcific deposits lying most often at the junctions of the bodies of the first and second sacral segments. Occasionally they can be seen also between the second and third sacral segments, but are usually smaller than the one above. The vertebral arches unite with the bodies of the lower sacral vertebræ at about two years of age, and of the upper segments at about six years. Epiphyseal plates for the superior and inferior aspects of the vertebral bodies appear at about sixteen years, and epiphyses appear on the lateral aspects of the sacrum at between the eighteenth and twentieth year.

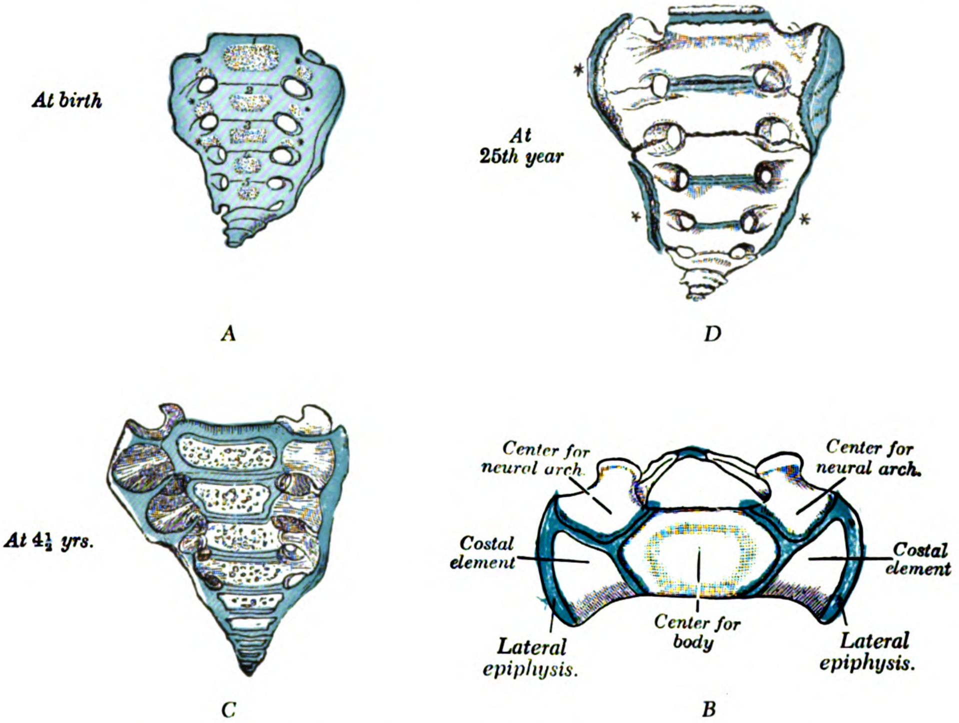


FIG. 14.—Ossification of sacrum. (Gray's *Anatomy*, Lea & Febiger.)

Each coccygeal segment ossifies from a single center. The first appears between the first and fourth year of life, the second at the fifth to tenth year of life, the third between the tenth and fifteenth year and the fourth between the fourteenth and twentieth year. Sometimes the segments may fuse with one another, and then may be identified only by a thin radiolucent line between them.

The vascular supply of the intervertebral disc is of considerable interest, and has clinical bearing on the explanation of changes which occur in these important structures. Very early in fetal development blood vessels extend to the intervertebral disc and regress equally rapidly. At about the third month of gestation, vessels appear which run parallel to the notochord. Others derived from the periosteum also penetrate into the cartilaginous plates without entering the central zone of ossification of vertebral bodies. These vessels enter the intervertebral cartilage at regular intervals along the rim of the vertebral body, and run in the direction of the nucleus pulposus. These radially placed vessels are responsible for the dentate appearance of the epiphyseal plates of the growing vertebræ and the deep indentations which are sometimes seen in the bony vertebral bodies of growing children. Zones of calcification and numerous foci of ossification may develop along these vessels, and these later in life fuse to form the ring apophyses. Still other vessels emerge from the vertebral body and penetrate into the cartilaginous plates directly, forming anastomotic arcades with those coming from the periosteum. Regression and thinning of the vascular supply to the intervertebral disc begins shortly after birth and continues progressively, so that by the age of eighteen to twenty five years most of the vessels have practically disappeared. Where these vessels have penetrated cartilaginous plates some gaps may result in the chondrification of the cartilaginous plates. At the time of complete degeneration of the vessels these chondrification cups may be replaced by scar tissue and sometimes by calcification. Their importance is that areas of diminished resistance appear to the increased turgor of the semi-solid intervertebral disc substance, particularly if the disc should be subjected to increased pressure. Through these points of reduced resistance nuclear prolapse occurs, forming so-called Schmorl's nodes. The nucleus pulposus receives no vascular supply directly.

The only source of nutrition for the tissues of the intervertebral disc is by diffusion processes through the cartilaginous plates coming from the cancellous vertebral bodies. The intervertebral disc presents its maximal elasticity at about the twenty-fifth to thirtieth year of life. Following this, the discs are subjected to the gradual processes of attrition, depending upon the strains to which the structures are subjected together with the chemical and hydropic constitutional alterations of the cartilage.

The nucleus pulposus of the intervertebral disc is derived from the notochord. Soon after the notochordal cells are squeezed into the disc, the nucleus pulposus grows more rapidly than the annulus fibrosis. The notochordal cells proliferate in local expansions in the discal centers while those in the vertebral bodies disappear. At about 6 months the discal notochordal cells begin to degenerate, and clump together to form a mucoid core surrounded by fibrous tissue and hyaline cartilage cells. Collagenous fibers extend into this gelatinous mucoid structure from the adjacent fibrocartilaginous capsule. In the primitive disc the nucleus pulposus is centrally situated, and the fibroblasts peripheral to it participate actively in the formation of the annulus fibrosus. The outer fibrous zone of the intervertebral disc is differentiated from the hyaline or precartilaginous inner zone. Notochordal cells can be demonstrated in the nucleus pulposus until adolescent life, and sometimes even later. The nucleus pulposus remains centrally situated in the cervical and thoracic regions, while in the lumbar discs it is somewhat more posterior. The fibrocartilaginous annulus fibrosus is heaviest anteriorly and laterally, while its posterior aspects are relatively weaker. Following the avascularization of the disc which occurs progressively until the third decade of life, the nucleus pulposus gradually is replaced by fibrous tissue, losing its gelatinous consistency. The question of innervation of the annulus fibrosus still is under discussion. There is some clinical evidence that these structures have a nerve supply, but until quite recently this has been denied.

The arterial supply to the vertebræ, the spinal ligaments and periosteum, and the spinal cord and its coverings is derived from branches of the vertebral, ascending cervical, deep cervical and occipital arteries in the cervical region, from dorsal branches of the intercostal arteries in the thoracic region and from posterior branches of the lumbar arteries in the lumbar region. The sacrum is supplied by lateral branches of the sacral arteries. Each spinal artery enters through its corresponding intervertebral foramen, and divides into three branches supplying the vertebral arches and ligamentum flava (postcentral branch); the spinal nerve, cord and adjacent membranes (neural branch); and to the posterior aspect of the vertebral bodies (the prelaminar branch). The latter also provides branches which extend to the anterior aspect of the spinal canal to anastomose with branches from a median artery on the dorsal surface of the anterior longitudinal ligament.

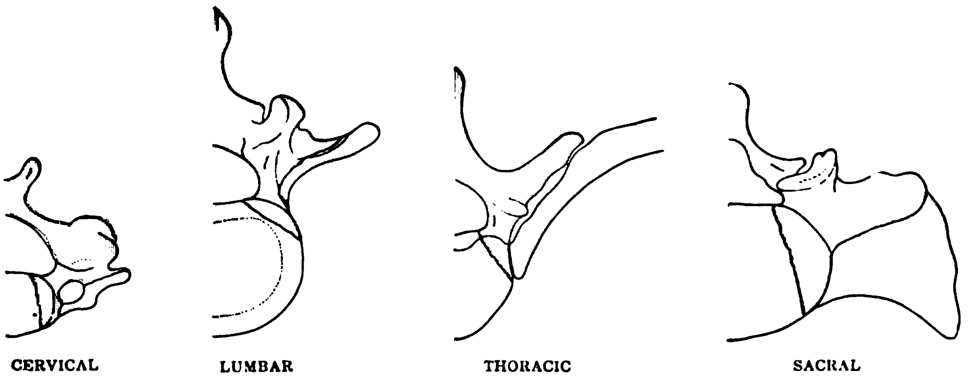


FIG. 15.—Diagrams showing the portions of the adult vertebræ derived respectively from the bodies (yellow), the vertebral arches (red) and the costal processes (blue). (Gray's *Anatomy*, Lea & Febiger.)

The venous supply of the vertebræ and spinal canal drains the spinal column, the meninges and adjacent musculature (Harris, 1941). This complex system is not accompanied by arteries, and is without valves. It is formed by large plexuses within and around the spinal column extending the entire length of the spine. The intraspinal plexuses course in the vertical direction as four large longitudinal veins, two in front of and two behind the spinal cord, forming anterior and posterior groups. The anterior internal plexus consists of large veins on the posterior surface of the vertebral bodies and the intervertebral fibrocartilages to either side of the posterior longitudinal ligament. The posterior internal plexuses are situated to either side of the midline in front of the vertebral arches and the ligamentum flavum. These communicate with the posterior external spinal plexus of veins by channels passing through these ligaments. A series of venous rings join the anterior and posterior internal plexuses. The internal and external vertebral plexuses anastomose freely with each other, terminating in the intervertebral veins which accompany the spinal nerves through the intervertebral foramen. This system of veins was brought into clinical prominence by the work of Batson (1940, 1942), who clearly demonstrated its role in the spread of metastases to the brain without pulmonary involvement.

In their final forms the vertebral segments are composed predominantly of cancellous bone. Only a thin shell of compact cortex pierced by many small foramina for blood vessels envelopes the periphery of the vertebral bodies. The neural arches and the laminæ have a relatively greater amount of compact bone. The transverse processes and the spinous processes tend to have a greater proportion of spongiosa.

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FUNCTIONAL ASPECTS OF THE ARCHITECTURE OF THE SPINAL COLUMN

The human spine is composed of 33 vertebræ—7 cervical, 12 dorsal, 5 lumbar—and the sacrum consisting of 5 segments and the coccyx consisting of 4 or 5 segments. Counting the sacroiliac joints, the posterior intervertebral joints and the costovertebral joints, there are 97 diarthroses associated with the spine, each with its separate capsule and synovial system. All are paired except the joint between the atlas and the odontoid process of the second cervical vertebra.

The first and second cervical vertebræ form an articulation with the skull, to which they are attached by strong ligamentous connections. The occipital condyles articulate with the superior articular surfaces of the lateral masses of the atlas in an arrangement which permits a nodding motion of the head. The joint cavities of this articulation communicate with the joint between the odontoid process of the axis and the anterior arch and transverse ligaments of the atlas. This pivotal joint and the condyloid joints between the atlas and the axis permit nodding and rotary motions of the skull on the vertebral column. There is no intervertebral disc between the first and second cervical vertebræ, the first disc occurring between the second and the third cervical vertebræ. Successive intervertebral discs increase slightly and progressively in size to the lumbosacral articulation. Between the bodies of the first and the second sacral vertebræ one may frequently observe the remnants of an intervertebral disc. A similar structure is less often observed between the second and the third sacral segments. The remainder of the sacrum usually is firmly welded into one mass. The coccyx articulates with the sacrum, and its various segments usually are separated from each other by intervening discs of cartilage. However, fusion of one or more of coccygeal segments is not at all uncommon.

The ribs, which are attached to the vertebral column by strong ligamentous bands, present two articulations with the vertebral column; one between the heads of the ribs with the bodies of the vertebræ and the other between the necks and tubercles of the ribs with the corresponding transverse processes. These joints are of the gliding or arthrodial type. The first, tenth, eleventh and twelfth ribs each articulate with a single vertebra. The remaining ribs articulate with the vertebra above and beneath by two demi-facets in the posteroanterior and posterosuperior aspects of the vertebral bodies for the respective rib articulations.

The cervical vertebræ are the smallest of the true vertebræ, and are easily identified by their contours and the presence of a foramen in each transverse process. The first vertebral body is ring-like, and is made up of two lateral masses connected by an anterior arch and a posterior arch. The outer surface of the anterior arch is convex and presents a small tubercle in its mid-portion called the anterior tubercle for the attachment of the longus colli muscles. The inner aspect of the anterior arch is concave, smooth, and serves as a receptacle for the odontoid process of the axis. The posterior arch of the atlas presents a more convex and wider configuration. Its mid-portion is tipped by a small process called the posterior tubercle, which corresponds to a rudimentary spinous process. In its anterior and superior portion, a groove for the vertebral artery and the first cervical nerve may be identified. Its transverse processes present rounded distal tips, and are broader, heavier and longer than the remaining transverse processes of the cervical vertebræ, except for the seventh. The basal portions of the transverse processes are perforated by the foramina transversaria for the passage of the vertebral arteries and veins.

The lateral masses of the first cervical vertebra are rather heavy, and present superiorly concave oval articular surfaces for the occipital condyles. These surfaces sometimes are slightly constricted in their medial portions. The inferior surfaces of the lateral masses bear the inferior articular facets which articulate with the axis beneath. These articular facets are circular in form, flat or slightly convex, and are directed downward and medially. Their structure is such as to permit rotary movement of the head. A rather heavy ligament passes between small tubercles along the medial margin of each superior facet. This divides the vertebral foramen into two compartments, one for the odontoid process and the other for the spinal cord and its membranes. The vertebral canal at this site is quite large, and almost pentagonal in contour, sufficient to permit some lateral displacement of the atlas without necessarily compressing the spinal cord.

The second cervical vertebra, also referred to as the axis or epistropheus, is characterized by the presence of the odontoid process which extends upward from the superior surface of its body. The articular surfaces of the lateral aspects of the body of the second cervical vertebra lie rather horizontally, and present rounded, slightly convex articular surfaces directed laterally and upwards to support the corresponding surfaces of the inferior aspects of the body of the first cervical vertebra. The transverse processes of the second cervical vertebra are considerably smaller than those of the first, and are penetrated by a foramen transversarium on each side. The spinous process and the heavy lamina of the second cervical vertebra are inclined downward and posteriorly. The tip of the spinous process of the second cervical vertebra usually protrudes beyond the others, and not infrequently presents a bifid tip.

A constriction or neck exists at the junction of the odontoid process and the body of the second cervical vertebra until about the fourth to sixth year of life. This may be seen as a radiolucent band between the odontoid and the subjacent vertebral body. The anterior aspect of the dens is in fairly close proximity to the apposing articular surface of the anterior arch of the first cervical vertebra. Radiologically a space of 1 to 2 mm. between these two structures may be demonstrated.

The anterior aspect of the body of the second cervical vertebra slopes slightly anteriorly and terminates in a lip inclined downward following the contour of the rounded, sloping anterosuperior aspect of the body of the third cervical vertebra.

The seventh cervical vertebra may be distinguished from the other cervical segments by the presence of a distinctive elongated heavy spinous process. The transverse processes of the seventh cervical segment as a rule are somewhat more prominent than the others, and the foramen transversarium in this segment is smaller than those seen in the remaining cervical vertebra.

The remaining cervical vertebræ appear fairly similar one to another. The vertebral bodies are quadrilateral in configuration with rather curved angles. The anterosuperior aspects recess slightly in front of the more prominent anteroinferior aspects of these vertebral bodies. The articular facets are inclined more obliquely proceeding downward. The laminæ are practically overlapping and are heavy. The spinous processes are small but increase in size proceeding from the third to the seventh cervical vertebræ. Their tips may be bifid, or they may terminate in a bulbous or flattened fashion. The transverse processes are about even in size, and each is perforated by a foramen transversarium.

The thoracic vertebræ are larger in size than those of the cervical spine and smaller than those of the lumbar spine. They increase progressively in size from the first to the twelfth thoracic vertebræ respectively, and can be recognized by facets on their sides for articulation with the rib heads, together with articular facets on the transverse processes on all except the eleventh and twelfth segments. The pedicles and laminæ of the upper thoracic vertebræ are directed downward and posteriorly and practically overlap one another. In the lower thoracic spine the degree of imbricational overlapping of the pedicles and laminæ is less marked than in the upper region. The spinous processes are heavy, and those of the upper thoracic spine are directed obliquely and rather sharply downward. The spinous processes become more vertical in position proceeding down to the lower aspects of the dorsal spine. The transverse processes arise from the neural arches behind the superior articular processes of pedicles. They are directed laterally and backward, and each terminates in a blunt round tip. On their anterior aspects one may identify the small articular surface for the rib tubercle.

The first, ninth, tenth, eleventh and twelfth dorsal vertebræ vary from the others in several respects. On either side of the body of the first thoracic vertebra is an articular facet for the first rib, while on its inferior portion a demi-facet for the costal articular process of the second rib can be seen. The transverse processes of the first dorsal vertebra are quite long. The body resembles a cervical vertebra, being quadrilateral and lipped on its antero-inferior and anterosuperior portions. The ninth dorsal vertebra has a demi-facet on its superoposterior aspect only. The tenth, eleventh and twelfth dorsal vertebræ present one entire facet corresponding to the articular heads of the respective ribs. The transverse processes of these three vertebral segments are rather variable, but usually are short and present no articular facets.

The lumbar vertebræ are the heaviest of the spinal column, and are distinguished from the others by the absence of foramina transversaria or facets on the side of the body as well as by their distinctive size. As a rule, the vertebral bodies are wider from side to side than from before backward, and are a little thicker in front than behind. The pedicles are directed backward from the upper part of the vertebral body, so that the inferior vertebral notches are of considerable depth. The pedicles and laminæ are dense and strong. The vertebral foramina present a triangular configuration and are larger than those in the thoracic spine and smaller than those in the cervical region. The spinous processes are thick, broad, somewhat quadrilateral and project downward and backward. These end in rough uneven borders which sometimes are notched. The superior and inferior articular processes of the neural arches are well defined, projecting upward and downward respectively from the junctions of the pedicles and laminæ. The transverse processes vary in size; the first, second and fourth being somewhat shorter than the third. The fifth transverse processes are variable and will be discussed further when considering the anatomy of the fifth lumbar segment. In the first, second and third lumbar vertebræ, the transverse processes arise from the junctions of the pedicles and laminæ, but in the fourth and fifth lumbar vertebræ they are set rather forward and originate from the pedicles and posterior portions of the bodies. The transverse processes are considered homologous with the ribs.

The fifth lumbar vertebra is subject to many variations in structure. The most common configuration of this segment is one in which the body is deeper in front than behind. Its transverse processes, as a rule, are heavier than those of the suprajacent vertebræ, and in the completely normal subject present rather bulbous tips which do not articulate with sacrum or iliac bones. The articular facets between the fifth lumbar and the first sacral vertebræ are very variable even in the normal subject. The true vertical placement of the articular facets which is seen particularly well between the second and third and fourth lumbar vertebræ is quite a rarity in the lumbosacral region. In many instances these articular facets may be completely coronally placed while in others the articular facets may be obliquely situated. Asymmetry between the articular facets on the right and left sides is not at all uncommon. The clinical significance of these variations in the apophyseal joints between the fourth and fifth lumbar and the fifth lumbar and first sacral vertebræ will be taken up in a later chapter.

The neural arch of the fifth lumbar vertebra is often subject to variations which are probably congenital in origin, although a great amount of discussion about the causes for incomplete fusion of the laminæ of the fifth lumbar vertebra has taken place. In occasional cases similar changes may occur in the fourth and to a lesser degree in the neural arches of the other lumber vertebra.

The normal sacrum is composed of 5 fused vertebral bodies. It is wedge-shaped, with its apex directed downwards. Seen laterally, the sacrum is tilted dorsally, and is convex posteriorly. The degree of dorsal tilting is greater in females than in males. Occasionally, the first sacral segment may be incompletely fused, and this is referred to as lumbarization of the first sacral segment. In other instances there may be fusion of the body of the fifth lumbar vertebra with that of the sacrum, giving rise to 6 sacral segments, a condition often designated as sacralization of the fifth lumbar vertebra. To either side of the mid-line, both on the dorsal and ventral aspects, are the paired sacral foramina. Between them are transverse lines separating the various vertebral bodies. These are not visible on the dorsal aspect of the sacrum, and can be identified on the lateral studies only on the anterior surface of the bone. As a rule, 4 sacral spinous processes can be identified. Beneath the distal one and between the fourth anterior sacral foraminæ the inverted U-shaped sacral cornua can be seen between which the sacral hiatus can be identified. The 4 sacral foramina are roughly and somewhat irregularly circular in configuration when observed anatomically. Their posterior aspects in the dorsal plate of the sacrum are somewhat smaller than their anterior fellows. They diminish somewhat in size from above downward. Beneath the fourth sacral foramen a notch may sometimes be observed, corresponding to the fifth sacral foramen.

The coccyx articulates with the apex of the sacrum which presents a small oval flat surface formed by the body of the fifth sacral vertebra. The dorsal aspect of the first coccygeal segment presents two upward processes referred to as the coccygeal cornua. The coccygeal segments represent rudimentary vertebræ, as a rule, only the body being present. These decrease in size from above downward and terminate in a small, somewhat oval flat bone. The first and second, and the second and third coccygeal vertebræ are usually united by a layer of fibrocartilage. The third and fourth and fourth and fifth segments often are fused. Considerable variation in the configuration and position of the coccygeal segments is frequently observed.

Twenty-three intervertebral discs unite the vertebral bodies from the second cervical to the first sacral segments inclusive, forming a series of amphiarthrodial or slightly movable joints. No intervertebral disc is present between the cranium and the first and second cervical vertebræ. In adult life the intervertebral discs are avascular and are composed of a surrounding annulus fibrosus made up of hyaline fibrocartilage enclosing the nucleus pulposus, a semigelatinous ovoid tissue whose function is principally that of maintaining the elasticity and bending abilities of the spinal column. It has been estimated that approximately one-fourth of the length of the vertebral column is made up of the intervertebral fibrocartilages. The sizes of the intervertebral discs correspond to those of the intervening vertebræ, so that the upper cervical discs are small while those in the lower lumbar spine are correspondingly large. Each disc is separated from the cancellous bone of the vertebral body by a plate of hyaline cartilage which in the child fits over the body of the vertebra very much as an epiphysis. Growth in the vertebra takes place at these epiphyseal plates. At about the age of six years, a ring apophysis appears as a narrow cartilaginous round structure lying on the rims of the cephalic and caudal surfaces of the vertebral bodies. These take no part in the longitudinal growth and represent an apophysis of the traction type corresponding to the attachments of the longitudinal and intervertebral, ligaments of the spine. Calcification occurs in these rings separately, and they fuse with the vertebral bodies at about the age of eighteen years. Beginning at about the age of eight years, they are often present in the thoracic and lumbar regions as irregularly calcified thin, broken circular bands. Later the calcification becomes somewhat heavier until final fusion occurs.

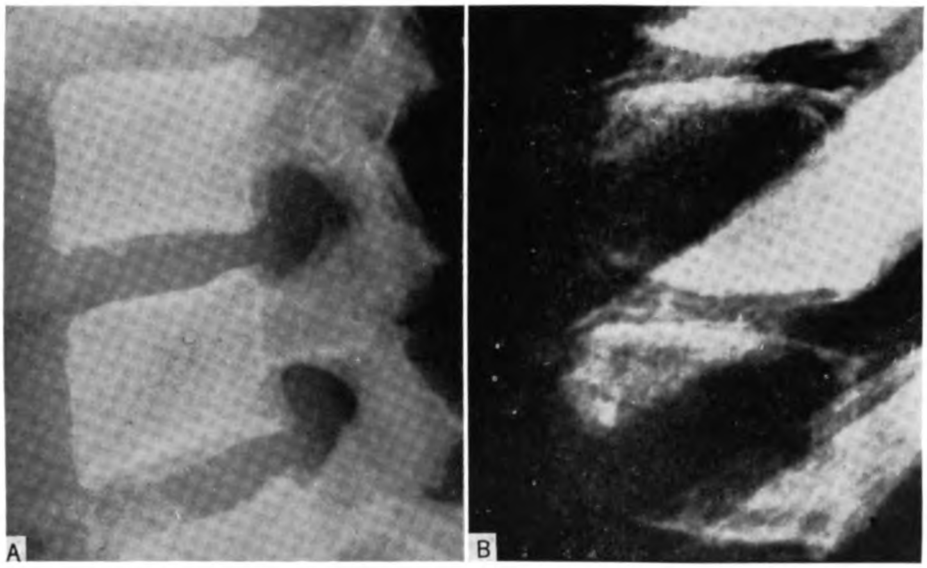

FIG. 16.—*A*, Lateral view of upper lumbar vertebræ of a fourteen-year-old girl showing ring apophyses. These are better formed in the thoracic vertebræ (*B*).

Vascular channels can be found in the cartilaginous intervertebral plates only during the first three decades of life. Later the discs are almost avascular except for some questionable channels in their peripheries.

In adolescent spines the intervertebral discs may be slightly expanded in the regions of the nucleus pulposus, but this recedes with life's progression of wear and tear. It has been estimated that in the average healthy young adult the gelatinous nucleus pulposus is under a pressure of 30 pounds to the square inch. The cartilage plates of the intervertebral discs not infrequently present small irregular breaks in their surfaces, and in children up to about the age of ten the cartilage plates may dip into the underlying cancellous bone in small grooves.

The annulus fibrosus is composed histologically of a mixture of fibrous cartilage and collagenous fibrous tissue arranged in concentric lamellæ passing obliquely across the intervertebral space in alternate directions. The fibers which arise as continuations of the fibrous cartilages of the apophyseal rings sweep around in an outward arch to be inserted in the opposite cartilaginous plate and bony ring. Some blend with the fibers of the longitudinal spinal ligaments and others turn over to become embedded in the outer cortex of the vertebral body on the side of the disc from which they originate.

The nucleus pulposus is composed histologically of a chondroid basophilic matrix traversed by a few delicate collagenous fibrils, sometimes containing stellate cells and at other times containing large vesiculated balloon cells supposed to represent vestigeal notochord. Notochordal remnants also have been identified in the vicinity of the first and second cervical vertebræ and the clivus Blumbenbachii.

As mentioned before, the spinal column is composed of two series of joints. These are the amphiarthroses or half joints formed by the inter-

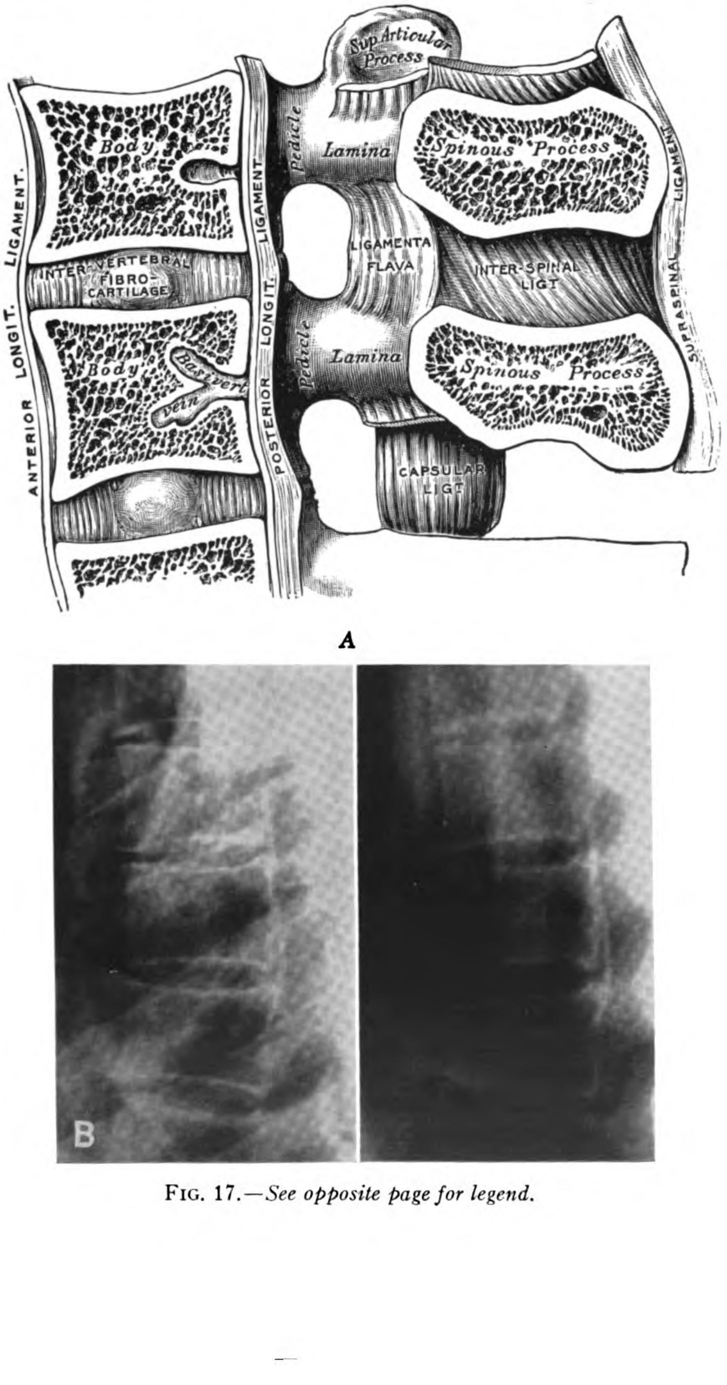

FIG. 17.—*See opposite page for legend.*

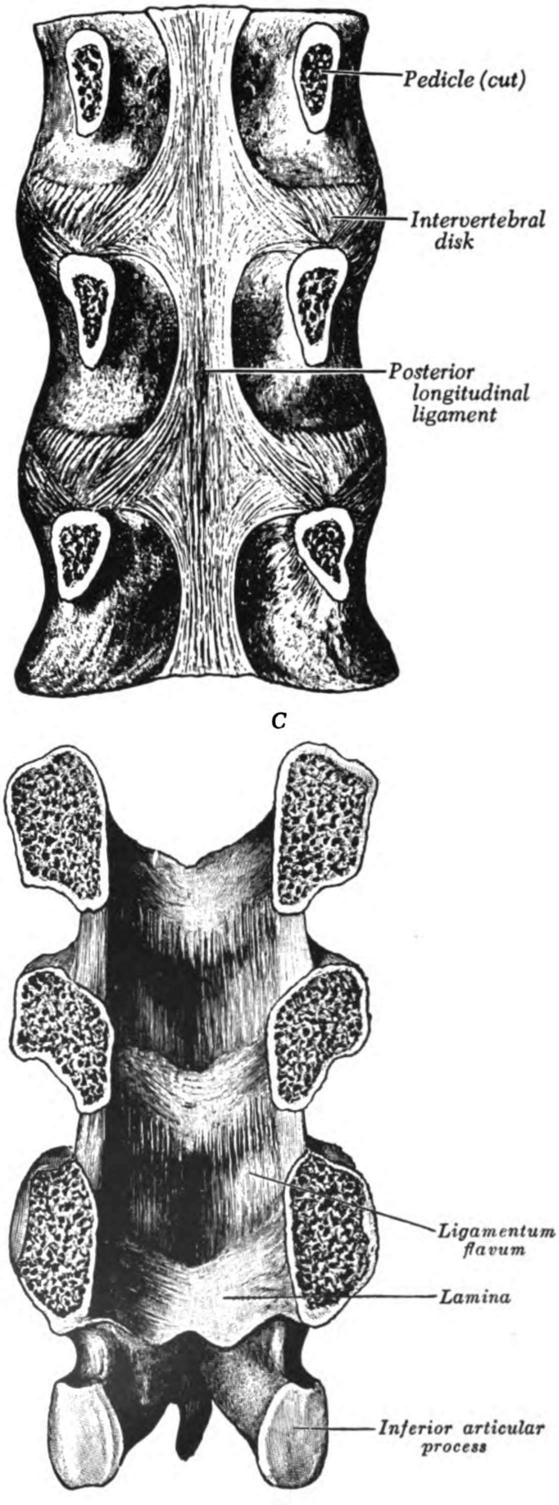

FIG. 17.—*A*, Medial sagittal section of two lumbar vertebræ and their ligaments. *B*, Lateral roentgenogram and laminagram showing calcification of the posterior longitudinal ligament in an adult asymptomatic patient. *C*, Posterior longitudinal ligament of the lumbar vertebræ. *D*, Anterior aspect of the lumbar ligamenta flava. (*A*, *C* and *D* from Gray's *Anatomy*, Lea & Febiger.)

vertebral discs, and the diarthrodeses, or true joints, containing synovial lining and articular cartilages in capsules formed by the inferior pair of articular processes, hooking down behind the superior segments below. These are designated as apophyseal, or posterior joints. Small joints are present at the posterolateral aspects of the lower five cervical vertebræ in the vicinity of the uncinate processes. Originally described by Luschka in 1858 as true synovial joints, they are important because their relationship to the intervertebral foramina is such that any overgrowth may impinge into the foramen and compress the contained nerve trunk. Generally they are considered true joints, but question as to the presence of a synovial lining has been raised at times, and certain authors regard them as degenerative alterations. Luschka's joints also are referred to as uncovertebral joints, covertebral joints or neurocentral joints.

Anteroposterior motion of each vertebra takes place about a transverse axis near the posterior border of its body. Flexion is checked by the supraspinatous, interspinous and interlaminal ligaments which bind the adjacent vertebræ together posteriorly. This is aided to some extent by the ligaments flava, which differ from the other ligaments in that they contain elastic fibers. These ligamentous bindings are so firm that except in the cervical area acute flexion trauma may result in compression of a vertebral body rather than rupture of the posterior check ligaments or dislocation of articular processes. Extension of the back is limited by the anterior vertebral ligaments and the intervertebral discs, which remain intact in extension injuries, the pedicles and lamina suffering fractures as a result of severe hyperextension.

At the cephalic end of the vertebral column, the intervertebral ligaments are represented by the heavy craniovertebral group. In the thoracic region the intervertebral ligaments are reinforced by the costo-vertebral ligaments, and at the lumbosacral region the strong lumbosacral, sacro-iliac and lumbo-iliac structures help stabilize the vertebral column on the pelvis. These groups of ligaments together with the inter-transverse group determine the degree of normal lateral motion and rotation of the spinal column.

The anterior spinal ligament is a heavy, broad band extending from the axis to the sacrum along the ventral aspect of the vertebræ. It is heaviest in its proximal portion, and is thicker where in apposition to the vertebral bodies. Its dense longitudinal fibers are fixed more firmly to the intervertebral fibrocartilages and the rims of the vertebræ than to their concavities. The ligaments at the sides of the vertebræ extend from these, and are considerably shorter and less dense. The posterior spinal ligament is situated within the spinal canal, extending from the body of the axis to the sacrum. It is heaviest in the thoracic region. This ligament is in intimate approximation to the intervertebral discs, particularly in their midportions. On either posterolateral side the ligament thins out, especially in the vicinity of the intervertebral foramina, where the discs bulge dorsally to occupy a variable portion of the lower half of the foramen. In the thoracic and lumbar regions the ligament follows the concavities of the respective vertebral bodies, but is thickened over the median line where it is separated from the bone by the basivertebral veins.

The ligamentum flavum, composed predominantly of yellow elastic tissue, joins the laminæ and articular processes as a continuous structure formed by a medial thicker half uniting the laminæ and a lateral thinner portion surrounding the articular joints and blending with their capsules. The interlaminar fibers are vertically disposed, and are attached inferiorly to the groove on the upper border of the inferior lamina and superiorly to the lower half of the superior lamina. Sharp thin spicules and plaques of bone extend into the ligament from its inferior attachment. These are continous ligaments, the inner thicker fibers being vertically arranged and the outer thinner ligaments are directed obliquely downward and outward. The ligament is so arranged that the inferior half of the lamina is excluded from the vertebral canal. Medially it blends with the opposite side at the bases of the spinous processes. The ligamenta flava are then broad and long in the cervical region, and increase in thickness caudally.

The muscles of the back are arranged symmetrically in opposing groups. These support the skeletal structures and control the motion of the back within the limitations set by the ligaments. Posteriorly the short and long intervertebral muscles are attached to the posterior and lateral processes. The vertebral-costal, vertebral-iliac and vertebral-sacral groups produce extension of the back, and with other coordinated groups, rotation and lateral bending. At the cephalic end homologous muscles control the motion of the head and neck. Altogether, the muscles form a heavy coordinated integrated mass making up the posterior wall of the torso. Flexion of the back is produced by contraction of the psoas muscles when the thighs are fixed, by the anterior and lateral abdominal muscles acting through the levers of the thoracic cage and pelvis, and by the anterior neck muscles. Rotation and lateral bending are accomplished by coordinated action of all groups (Willis, 1944).

At about the third month of gestation, the fetal spine assumes a single dorsally convex somewhat coiled posterior curve. Uncoiling begins between the third and sixth months, when the movable portions of the vertebral column straighten out and acquire a second ventral curve. At birth, a posterior thoracolumbar convexity and a sacral curve are present. The head, hips and knees are flexed, and when the face is tipped upwards the anteriorly convex cervical curve develops. When the infant's hips are extended the lumbar vertebræ are pulled forward by the psoas muscles and hip ligaments forming a typical lumbar curve. This does not become firmly fixed until the child has been walking for several years. In the adult, the cervical spine in the resting position presents a normal anterior curve. The dorsal spine then has a slight dorsal curve and sometimes a curve convex towards the patient's right side. The usual lumbar lordotic curve is demonstrable in the lateral position. This increases somewhat when the patient is in the erect posture.

The vertebral canal follows the various curves of the spinal column. The canal is large and somewhat triangular in the cervical and lumbar portions of the spine where the degree of movement is greatest. In the thoracic region, where motion is restricted, the spinal canal is relatively narrow and circular.

Two types of curves, the primary or accommodation curves, and the secondary or compensation curves are encountered in the spinal column. Four of these exist in the sagittal plane, convex forward in the cervical and lumbar regions and concave forward in the dorsal and sacral ones respectively. The dorsal and sacral curves are primary, persisting from the embryonic state. These accommodate the thoracic and pelvic viscera. The cervical and lumbar curves are convex ventrally and represent secondary curves that appear when the upright posture is attained, and develop by changes in the intervertebral discs. The cervical curve appears in infancy when ventral flexion of the head on the chest begins at about the third month, and with ability to sit at about the sixth month. This cervical curve is not fixed, and is present in the upright but obliterated in the horizontal posture. The lumbar curve between the twelfth thoracic vertebra and the sacral promontory appears between the ages of nine and twelve months, when the child begins to stand and to walk. This curve becomes relatively fixed in adult life. The anterior prominence of the cervical curve is situated at the body of the fourth cervical vertebra, and the curve ends at about the second dorsal vertebra. The most prominent portion of the dorsal curve is at the seventh or eighth dorsal vertebra, and its termination is at the twelfth thoracic or first lumbar vertebra. The fifth curve occurs in the dorsal spine as a lateral secondary or compensation curve. In most cases it is directed toward the right, and is probably associated with the greater use of the right hand. This is often referred to as physiologic right lateral scoliosis. The lateral curves of accommodation have associated minor morphologic changes in the bodies of the dorsal and sacral regions, while the secondary or compensation curves have associated changes at the intervertebral discs.

A variable flattening of the left side of the dorsal vertebræ from the fourth to tenth dorsal segments inclusive may exist, forming the so-called aortic compression.

The alternating curves of the vertebral column are largely responsible for the strength of the spine, which is estimated as sixteen times that of a straight alignment. The curves are so situated as to receive thoracic and abdominal organs.

The intervertebral foramina are oval in shape, and increase in size from the cephalic to caudal regions. By virtue of the movement of the vertebræ they accommodate themselves to the various motions of the spine. When the neck is flexed the intervertebral spaces are oval and fairly large, but in extension the foramina decrease in size as the gliding surfaces of the articular facets pass one over the other as inclined planes. In the thoracic spine, the intervertebral foramina are relatively small and contain little adipose tissue, since little cushioning of the nerve roots is required because of the restricted motion of the respective dorsal segments. In the lumbar region, posterior extension of the spine causes relative constriction of the intervertebral foramina, while anterior flexion increases the diameter of these openings. It has been estimated that the spinal nerves occupy about one-quarter of the available space of the respective foramina, the rest being utilized by fatty tissue, vessels and lymphatics.

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THE SPINAL CORD

The embryologic derivation of the spinal cord is from the neural groove, a structure of ectodermal origin from which the nervous and neuroglial elements of the spinal cord appear. The central canal of the spinal cord persists as the rudiment of the embryologic neural canal communicating with the cerebral ventricles in the same fashion that the spinal cord is a direct extension of the brain.

The coverings of the spinal cord are composed of the dura mater, the arachnoid and the pia mater. These originate from the primitive mesenchymal tissue surrounding the nervous system, and are derived from the same tissues which form the bone and periosteum. In the embryo the dura is separated from the nervous system by loose mesenchymal tissue containing albuminous tissue fluid. The pia mater is formed by mesenchymal cells on the surface of the neural tube. At the onset of secretion of the cerebro-spinal fluid, excess fluid passes into the tissue spaces separating the two leaves of the arachnoid into the subarachnoid spaces. Arachnoid trabeculæ extend from the pia to the peripheral portion of the dura. The dura and arachnoid are separated by a thin space containing a small amount of colorless fluid, presumably of lymphogenous nature.

The spinal cord originates as an open groove which later closes to form a tube. The distal portion of the spinal cord, however, develops differently in that it starts as a solid cord of cells rather than a closing or closed neural tube. The central canal in this area is present as a secondary structure rather than as a result of the closing of the original neural groove. Up to the third month of fetal life the spinal cord is about the same length as the spinal canal. During the subsequent growth of the fetus, there is a discrepancy in the growth of the spinal canal as compared with the spinal cord. The filum terminale then appears and inserts itself into the sacrococcygeal region. In the adult the filum terminale consists of an intradural portion about 1 cm. long and an external portion about 12 cm. long extending below the caudal sac and inserted in the first coccygeal segment. At birth, the lower pole of the spinal cord, as a rule, is located at approximately the level of the third lumbar vertebra, while in the adult it usually reaches the level of the superior margin of the second lumbar vertebra.

During the early stages of development, the dura mater is in close proximity to the spinal canal. Later these structures separate and the epidural space is formed. This space becomes filled with areolar tissues and vessels, predominantly large veins. The dura is attached to the intervertebral discs in the lumbar region. In the sacral areas the dura is separated from the anterior wall of the spinal canal and merges itself with the filum terminale and fastens with this structure to the anterior part of the sacral canal. Considerable variation is seen in the configuration of the distal end of the caudal sac, and individual differences are noted in its point of termination, which usually is at approximately the level of the fifth lumbar intervertebral disc.

The spinal dura mater is fixed to the circumference of the foramen magnum and the second and third cervical vertebræ. It represents only the inner or meningeal layer of the cranial dura mater. The outer or endosteal layer of cranial dura mater terminates at the foramen magnum and is replaced by the periosteum lining the vertebral canal. The dura is connected to the posterior longitudinal ligament, especially at the distal portion of the vertebral canal by fibrous tissue. The dura is quite loose and permits ample space, particularly in the cervical and in the lumbar regions. In the dorsal region, however, the dura is more closely affixed to the spinal canal and the intradural space is considerably less than in other areas. On either side of the dura are the extensions which invest the corresponding spinal nerves. These are short and relatively horizontal in the uppermost portion of the spinal canal, particularly in the cervical and upper dorsal vertebræ. As the cord proceeds downward, they become gradually longer and are directed obliquely downward, extending in some instances into the vertebral canal for varying distances. The dura is only poorly supplied with blood vessels and contains a relatively scant supply of nervous tissue.

The arachnoid coat of the spinal cord is composed of tubular delicate membranous tissue continuous above with the cranial arachnoid. In its distal portions, it surrounds the cauda equina and the various nerve roots forming this structure. The arachnoid coats the spinal nerves in loose sheaths and extends with them as far as their points of exit from the spinal canal. The subarachnoid space in which the cerebro-spinal fluid circulates is that area between the arachnoid and the pia mater. This space is occupied by delicate connective tissue in a spongy conglomeration, permitting many intracommunicating channels. It is in direct communication with the subarachnoid space of the brain and the ventricular system. On either lateral aspect of the spinal cord are the dentate ligaments, composed of a narrow fibrous band extending through the entire length of the spinal cord. These separate the anterior from the posterior nerve roots and are fixed into the pia mater in broad triangular bands and terminate in the opposite arachnoid in sharp, tooth-like processes, fixed at intervals to the dura mater. There are 21 dentate ligaments on either side, the first being attached to the dura opposite the margin of the foramen magnum between the vertebral artery and the hypoglossal nerve and the last situated near the distal end of the spinal cord.

The pia mater is a thin delicate vascular membrane formed by areolar tissue, and in the spinal canal consists of two layers. The outer one is composed of bundles of connective tissue fibers arranged in a longitudinal fashion, while the inner is approximated to the cord in a more circular manner. Between these two closely approximated layers of the spinal pia mater are other spaces which are in continuity with the subarachnoid cavity. This contains a rather extensive collection of blood vessels enclosed in perivascular lymphatic sheets. The pia closely envelopes the spinal cord and extends outward along the nerve roots as well. Along the midline of the anterior aspect of the cord a longitudinal fibrous band termed the linea splendens can be demonstrated. This is somewhat similar to the ligamentum denticulatum on either side of the spinal cord.

The pia mater terminates below the conus medullaris as the filum terminale, which extends downward to form the center of the mass of nerves

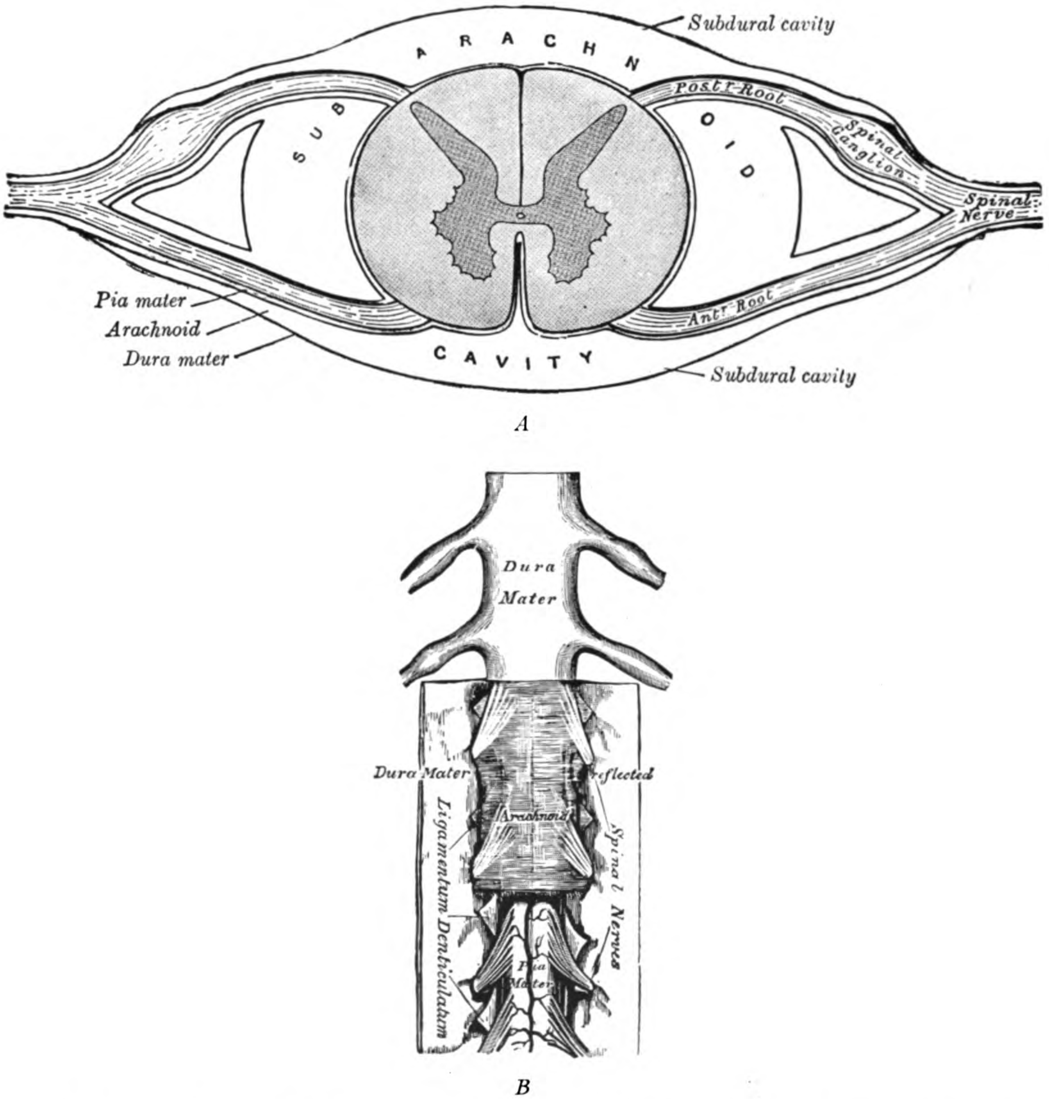


FIG. 18.—*A*, The dura mater and the arachnoid membranes. *B*, The spinal cord and its membranes, diagrammatic cross section. The dentate ligament, arising from the pia mater, is seen in *B*, and not represented in *A*. (Gray's *Anatomy*, Lea & Febiger.)

forming the cauda equina. The filum terminale blends with the dura mater at the level of the second sacral vertebra and then proceeds downward as far as the coccyx, where it fuses with the periosteum.

There are 31 pairs of spinal nerves arising from the spinal cord. Each possesses anterior or ventral and posterior or dorsal roots, the latter being identified by the presence of spinal ganglions. There are 8 pairs of cervical nerves, 12 pairs of thoracic nerves, 5 lumbar nerves, 5 sacral nerves and 1 coccygeal nerve. The first cervical nerve passes between the occiput and the first cervical vertebra, and the eighth emerges from between the seventh cervical and the first thoracic vertebræ. The first thoracic pair of nerves goes through the foramina between the first and second thoracic vertebræ, and so on downwards. Thus, the nerves below the cervicothoracic junction have the numerical designation of the vertebra immediately cephalad, as, for instance, the fifth lumbar nerve coming out from between the fifth lumbar and first sacral vertebræ. Even though there is no anatomic evidence to point towards specific segments in the spinal canal, it has become customary to divide the cord into spinal segments or neuromeres, each of which possesses length equal to the extent of the attachment of the corresponding pair of spinal nerves. These vary in various portions of the spinal canal. In the cervical region the neuromeres average about 13 mm. in length, in the midthoracic region about 26 mm. in length, while in the lumbosacral regions they diminish rapidly from 15 mm. at the level of the first lumbar nerves to about 4 mm. opposite the attachment of the lower sacral nerves. The configuration of the nerve roots vary considerably in relation to their situation within the spinal canal. In the uppermost portions of the canal the nerve roots are short, relatively fixed in position and emerge almost horizontally. In the lumbar and sacral areas, the nerves are inclined progressively downward so that in the distal portions of the spinal canal they emerge almost vertically to reach the intervertebral foramina. In the lumbar portion of the spinal canal the appearance of the spinal nerves has given rise to the descriptive term cauda equina.

The spinal cord itself is rather oval and flattened dorsoventrally, and is relatively larger in the cervical and lumbar regions than in the thoracic region. The cervical enlargement which is the more prominent extends from approximately the level of the third cervical vertebra to the second thoracic vertebra, reaching its widest diameter at the sixth cervical segment. The lumbar bulge begins at approximately the level of the ninth thoracic vertebra and terminates opposite the twelfth thoracic vertebra. Below this the cord tapers abruptly forming, the conus medullaris and the filum terminale.

INTRASPINAL DIMENSIONS.

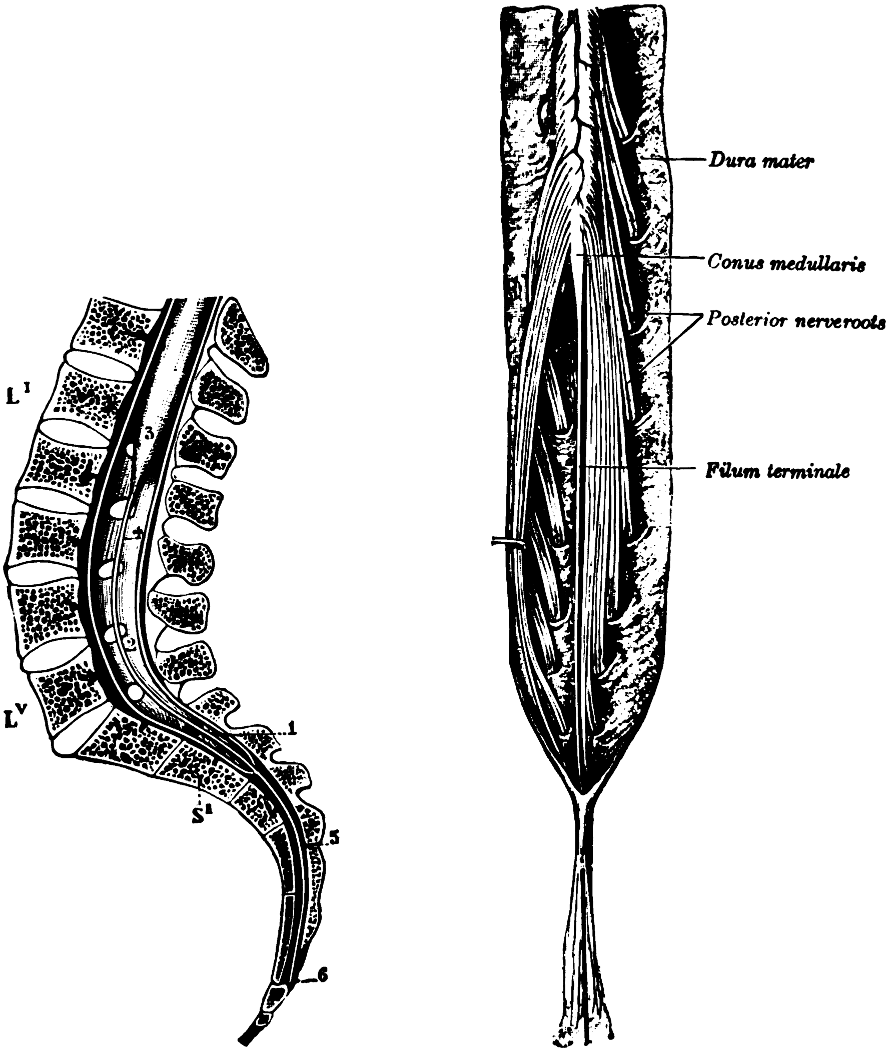
The volume of the spinal cord is difficult to evaluate accurately because it changes with varying conditions. During strain vascular dilatation produces diminution in the subarachnoid space, displacing cerebrospinal fluid during the period of vascular engorgement. Alterations in the alignment of the vertebræ also causes corresponding impingement into the spinal canal. The relative position of the spinal cord varies during motion, and with the assumption of various body postions. Arnell (1948) described the apparent descent of the cord when the individual is recumbent, and its approach to the dorsal aspect of the dural sac in the supine position. A lesser degree of displacement of the cord from side to side occurs when the right or left lateral recumbent position is assumed. Flexion and extension

TABLE 1 — MEASUREMENT OF THE NORMAL INTERPEDICULAR SPACE IN THE CHILD.
(From Landmesser, and Heublein, courtesy of Connecticut State Med. J.)

	Vertebræ																
	Dorsal												*Lumbar*				
Age	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**	**1**	**2**	**3**	**4**	**5**
1	16–20	14–19	13–17	13–17	13–17	13–17	13–17	14–17	14–17	14–17	14–17	15–19	15–20	17–20	17–20	18–22	19–23
2	17–21	14–19	14–17	14–16	14–16	14–16	13–17	13–17	14–19	14–19	15–19	15–20	17–21	18–21	18–22	19–22	21–26
3	17–22	16–19	14–18	14–17	14–17	15–17	15–17	15–18	15–18	16–19	16–20	18–22	19–22	19–23	19–23	20–25	20–27
4	19–23	16–21	15–19	15–19	15–19	14–19	14–19	15–19	15–19	15–19	16–21	19–23	20–23	19–23	19–23	19–25	23–28
5	20–24	17–21	15–20	15–19	15–19	15–19	15–19	15–19	15–20	15–20	18–20	18–24	18–24	19–25	19–25	23–26	24–29
6	20–24	16–21	16–19	15–19	15–19	15–18	15–19	15–20	15–19	16–20	17–21	19–24	20–24	20–26	21–26	21–28	22–29
7	20–26	17–23	15–21	15–20	15–19	15–19	15–19	15–20	15–20	15–20	17–21	19–24	21–25	21–26	22–27	23–29	25–32
8	19–26	16–22	15–20	15–19	15–19	15–19	15–20	15–21	15–21	16–21	17–22	19–24	20–25	21–25	22–27	23–27	26–32
9	20–26	17–21	16–20	16–20	16–19	15–19	15–19	15–20	15–20	16–21	17–21	20–24	21–25	22–26	23–27	23–27	25–31
10	23–26	18–22	17–21	16–20	15–19	15–19	15–20	15–21	15–21	17–21	18–21	19–25	21–26	23–26	23–27	24–29	26–32
11	21–25	18–22	16–21	16–21	16–20	16–19	16–20	16–21	16–21	17–21	18–22	21–25	22–26	22–26	23–27	24–28	24–32
12	19–26	17–23	15–21	15–20	15–20	15–19	15–19	16–20	16–21	17–21	17–22	21–25	21–26	22–26	23–27	24–29	26–32
13	21–26	18–23	16–21	15–21	15–20	15–20	16–20	15–21	15–21	15–21	16–22	18–25	20–27	21–27	22–29	23–30	25–32
14	22–27	18–23	17–21	16–21	16–20	16–20	16–20	16–20	16–21	17–21	17–23	20–25	22–26	22–27	22–28	23–29	28–35
15	22–27	18–23	17–21	16–20	16–20	15–20	15–20	15–21	15–21	16–22	18–24	21–26	23–27	23–27	24–28	24–30	26–34

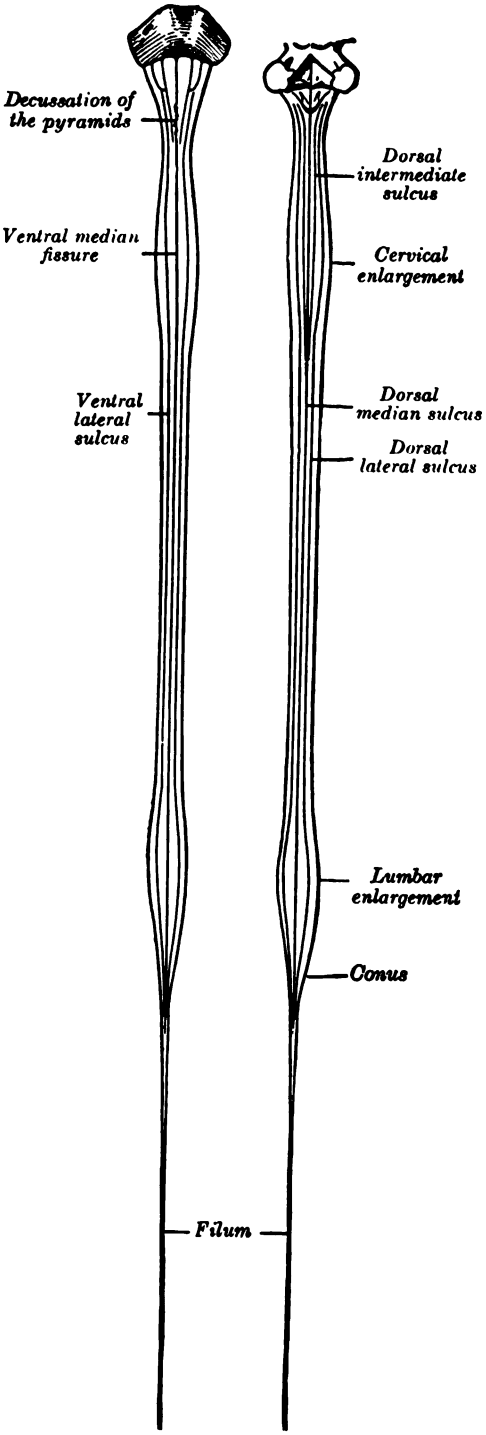
of the head and trunk stretch all segments of the spinal cord, most prominently in the cervical region and least in the caudal area. A lesser degree of change in position occurs during flexion of the head and trunk, and elongation of the cord takes place through traction on its nerve roots during movement of the extremities.

The subarachnoid space varies according to its situation in the spinal canal. In the cervical area the dural sac follows the cord contours rather closely, being flattened dorsoventrally. In the thoracic spinal canal the dura assumes an almost circular configuration, and as it invests the cord it is smaller than in either the cervical or the lumbar area. In the lumbar spinal canal the dural sac is triangular in the cephalad segments and almost pentagonal in the last two caudad vertebræ, with rounded angles following the general configuration of the region. The sacral dural envelope again becomes almost circular, but considerable variation in size and contour occurs here.



A B

Fig. 19



C

FIG. 19.—*A*, Vertebral canal showing lowering end of medulla spinalis and the filum terminale. *B*, Cauda equina and filum terminale, see from behind. *C*, Diagrams of the spinal card indicating positions of cervical and lumbar enlargment. (*Gray's Anatomy.*)

The dimensions of the cervical spinal canal in the sagittal plane vary from between 16 and 30 mm. at the first cervical vertebra. The second cervical segment averages between 14 and 28 mm., and the third from 12 to 24 mm. The remaining segments vary from 12 to 22 mm. in depth. The average values are 22 mm. at the first cervical vertebra, 20 mm. at the second and about 17 mm. from the third to the seventh cervical vertebræ. The subarachnoid space in the lower cervical canal anteriorly is approximately 6 mm. and posteriorly 4 mm. The thoracic spinal canal is approximately 17 to 22 mm. in diameter, and the subarachnoid space is approximately 8 mm. anteriorly and 4 mm. posteriorly. The lumbar spinal canal varies from approximately 18 to 25 mm. in its sagittal diameter for the upper there lumbar segments. Below this it diminishes somewhat to approximately 20 mm. In the upper lumbar canal the subarachnoid space is about 8 mm. anteriorly and 4 mm. posteriorly. Beneath this is the cauda equina, where the sagittal depth of the subarachnoid space varies from about 16 to 23 mm. (Arnell, 1948).

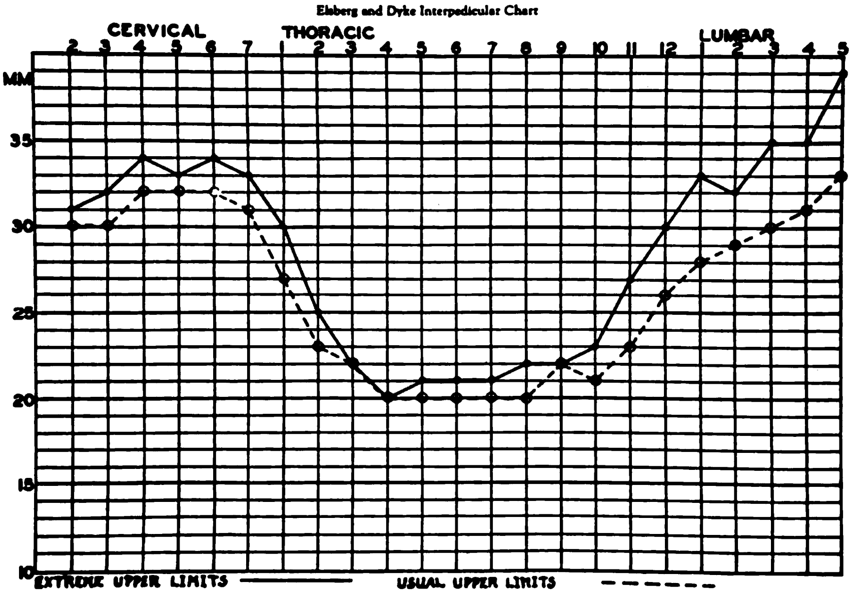


FIG. 20.—Elsberg and Dyke interpedicular chart.

The normal range of interpedicular measurements in adults was established by Elsberg and Dyke (Fig. 20). Similar values were determined for infants and children by French and Peyton (1942), who published two sets of curves for infants from birth to five years and from five to ten years of age. A more detailed survey was made by Simril and Thurston (1955), who found that three groups could be established, from birth to two months, from two months to two years and from two to twelve years. Landmesser and Heublein published a convenient table of measurements in children from one to fifteen years of age (Table 1).

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RADIOLOGIC EXAMINATION OF THE NORMAL SPINE

The Cervical Spine

The technic for radiographic examination of the cervical spine is somewhat different in the infant and young child as compared with the adult. In infancy, the probability of disease of the articulations is not very great. Here one is more concerned with anatomical details of structure. The technical aspects of achieving a sharply detailed study of the cervical spine in an infant or child is more difficult than the corresponding examination

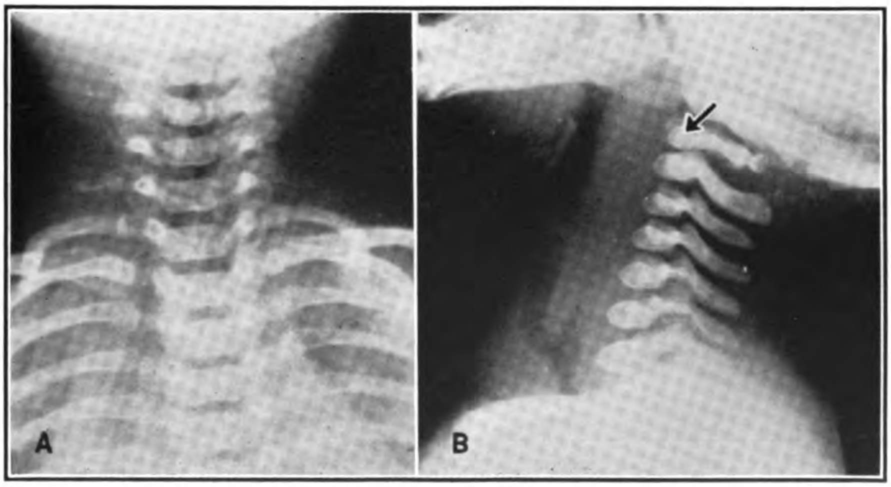


Fig. 21.—*A*, Anteroposterior view of cervical spine of newborn infant. Two faint centers of ossification are present in each vertebral body on either side of the midline. *B*, Lateral view, same case. Note the tendency to rounding of the anterior aspects of the cervical vertebral bodies. The ossification center of the dens (arrow) is not yet united with the body of the second cervical vertebra.

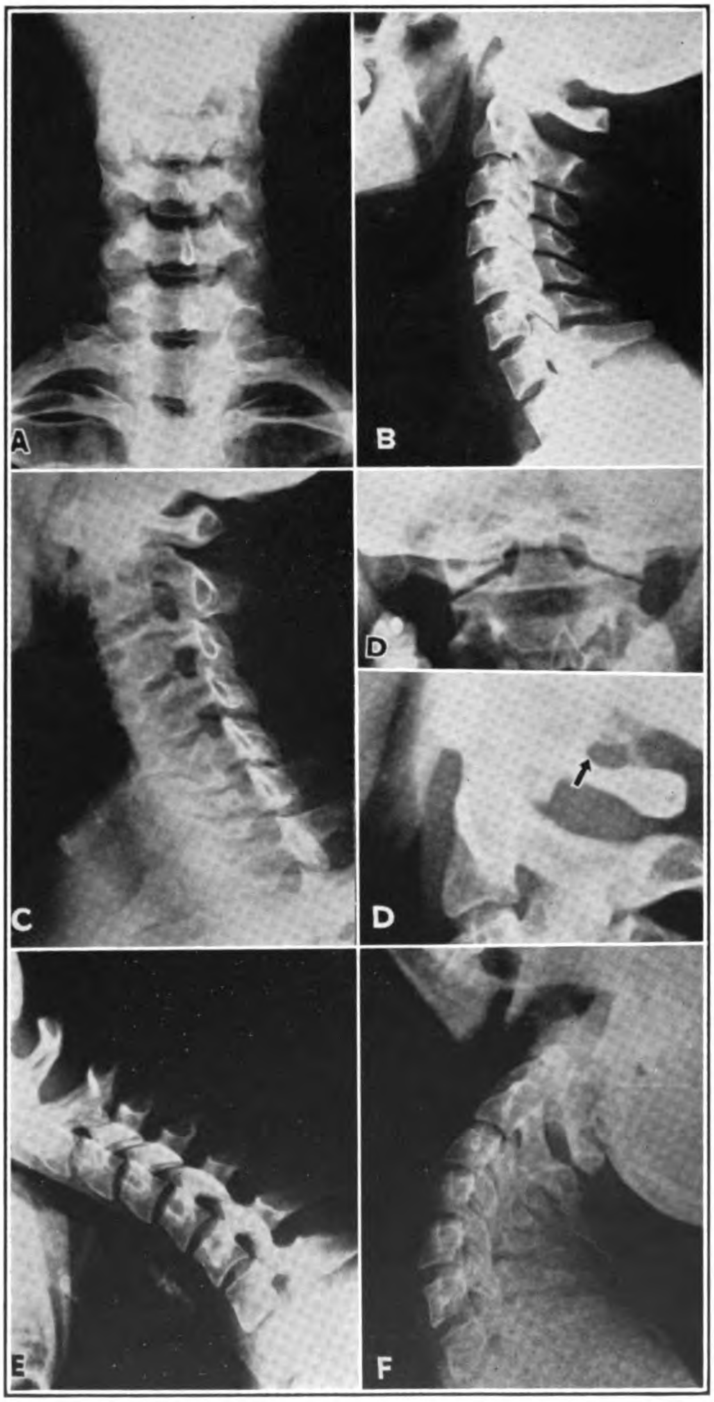


FIG. 22.—*A*, Anteroposterior view, normal cervical spine, adult. *B*, Same patient, lateral view, neutral position. Note normal lordotic curve. *C*, Same patient, right anterior oblique view demonstrating intervertebral foramina. *D*, Anteroposterior and lateral views of atlas and axis. Note the "ponticulus posticus" of the first cervical vertebra (arrow). *E*, Same patient, extreme flexion. Note gliding movement of articular facets and separation of tips of spinous processes. *F*, Same patient, extreme extension showing normal range of mobility.

in the older child or adult. It is our custom to examine the cervical spine in infants in the recumbent positions, in the anteroposterior and lateral views. In the lateral projection it has been found helpful to extend the child's neck manually, and simultaneously to bring the arms as far dorsally as possible with the shoulders drawn down. In this position, the entire cervical spine and sometimes the upper dorsal segment are brought into

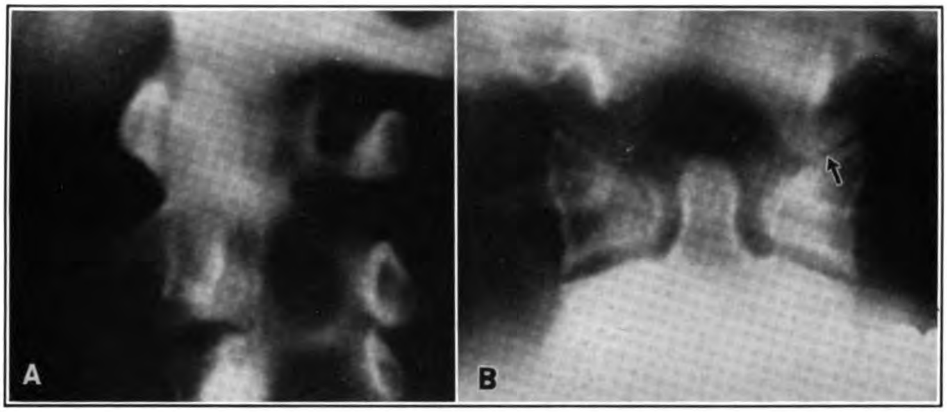


FIG. 23.—Lateral (*A*) and anteroposterior (*B*) body section roentgenograms of first and second cervical vertebræ. Note articulation of atlas with cranial occipital condyles (arrow).

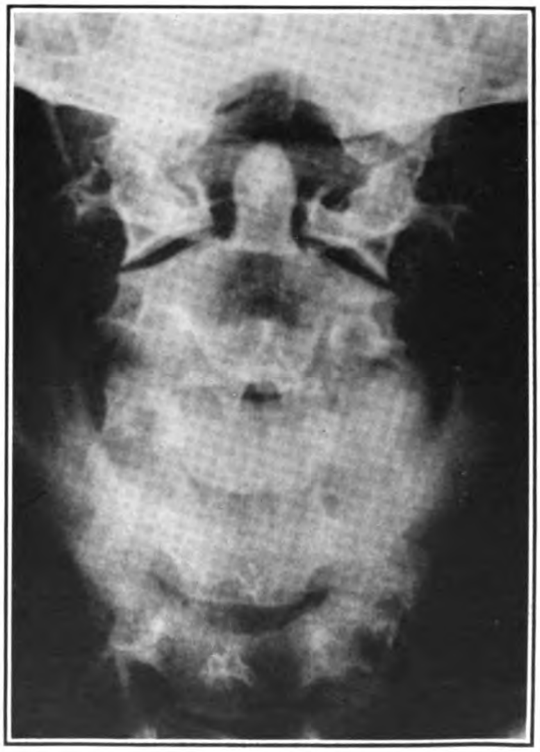


FIG. 24.—Anteroposterior view of upper two cervical vertebræ made with jaw in motion (Ottonello's technic).

view. The study is best made with an extension cylinder cone using $\frac{1}{60}$ of a second if possible, without the Bucky diaphragm, using screens of the finest definition. Unless specifically required for some unusual possibility, studies in the oblique positions are not required, nor is it often necessary to obtain flexion and extension studies.

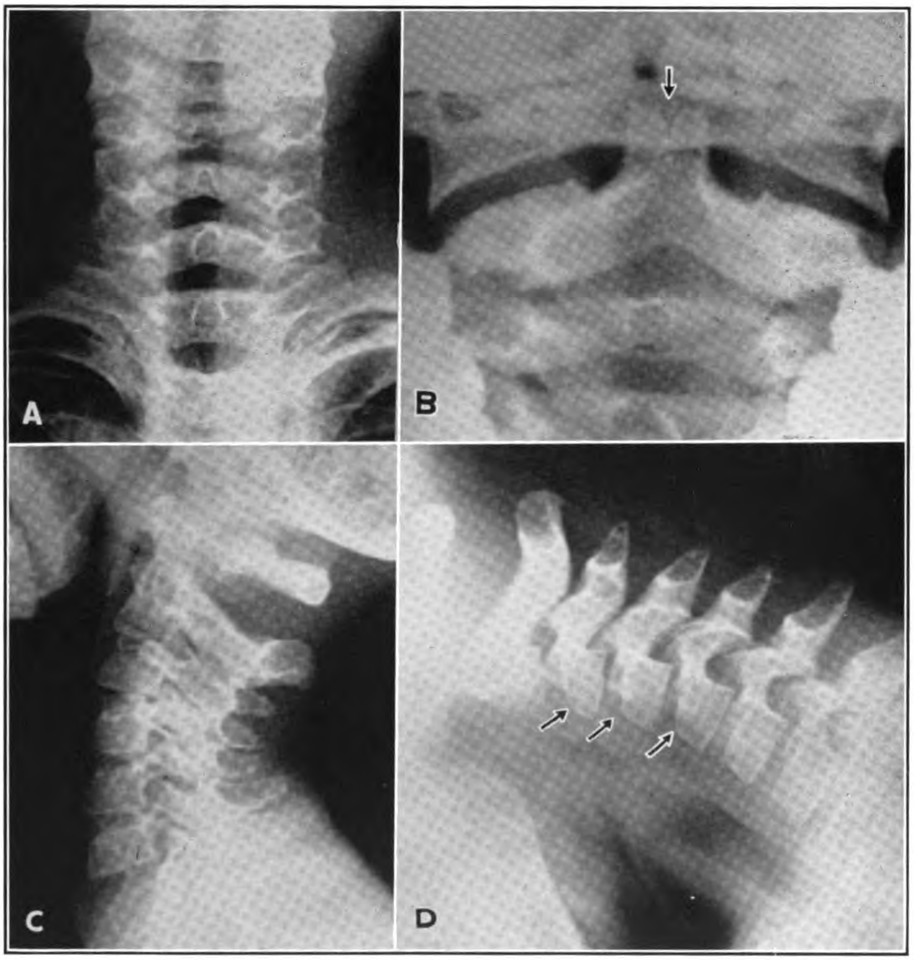


FIG. 25.—*A*, Anteroposterior view of cervical spine of normal five-year-old child. *B*, Same patient, atlas and axis. Note cleft in dens, with small center of ossification in midline (arrow). *C*, Same patient, lateral view. *D*, Same patient, lateral view in flexion. Note range of motion of vertebral bodies and their spinous processes. The anterosuperior aspects of the bodies are rounded (arrows).

In young children, adolescents and adults, the technic of examination of the cervical spine may be varied according to the information required. Study must be directed towards visualization of the vertebral bodies, the intervertebral spaces and foramina, the apophyseal joints, the transverse and spinous processes and the paravertebral soft tissues. One should include anteroposterior views, special studies for the atlanto-axial articulation, right and left oblique views and lateral views in the neutral position, in flexion and in extension. This, of course, represents optimum requirements and a degree of clinical judgment should be exercised in the selection of those projections required to answer a specific question.

Inasmuch as the present volume does not undertake to present radiologic technic in detail, no attempt is made to provide technical factors here or in subsequent sections. Suffice it to say that the interpretation of roentgenograms of the spine rests upon the firm foundation of proper technic, and films that are poorly exposed, made with the patient in motion or in improper position all may lead to diagnostic error.

The importance of obtaining stereoroentgenographic views is often insufficiently recognized. These studies provide a three-dimensional index for the separation of bony shadows and it is felt that the disadvantages of additional time and expense is well compensated by the additional information which one obtains from stereoroentgenograms.

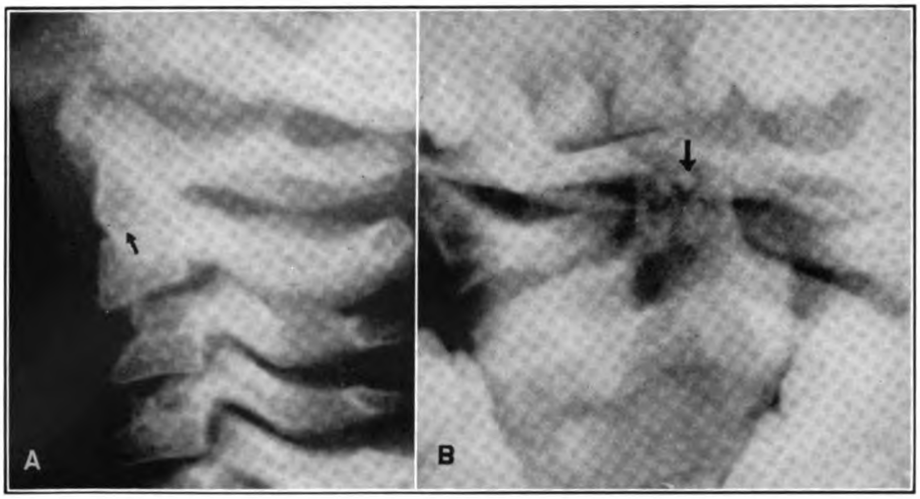


FIG. 26.—*A*, Normal atlas and axis, six-year-old child. Note line of junction of dens with body of second cervical vertebra (arrows). *B*, Same patient, anteroposterior view through open mouth. Note incomplete fusion in midline of dens, with small ossification center in cleft at its tip (arrows).

Laminagraphic examinations of the cervical spine have also proved helpful in visualization of the atlanto-axial articulation and visualization of the first and second cervical segments, particularly the odontoid process. The visualization of the atlas and axis may be obtained more satisfactorily employing a technic in which the mandible is kept in constant motion during the roentgenographic exposure, (Ottonello, 1930), the principle being that continued motion of the mandible during a rather long exposure eliminates the mandibular shadow, leaving the upper cervical spine in sharper relief.

In viewing roentgenograms of the cervical spine in newborn infants, one should recall that the odontoid process is cartilaginous and hence not visible. Its absence should cause no doubt. In later infancy and early childhood, as the odontoid begins to calcify, its structural characteristics become manifest. These are well seen as a ventral, split bar in the second and third year of life delineated from the anterosuperior aspect of the body of the second cervical vertebra by a thin line of cartilage. The tip of the odontoid is cleft by a small V-shaped defect, which later is occupied by a calcified epiphysis which ultimately fuses with the mass of the odontoid. The vertical cleft between the two lateral masses of the odontoid may occasionally persist into adolescence, while the cartilage line between the base of the odontoid and the subjacent body of the second cervical vertebra may be visible up to about the age of ten years. This sometimes may be mistaken for a fracture line, particularly when observed in the anteroposterior view through the open mouth.

The articulations between the occipital condyles and the first cervical vertebra can best be demonstrated in the anteroposterior position by means of laminagrams. These cup-like articulations can be well demonstrated on films made with the patient's mouth open. The superior facets of the body of the first cervical vertebra can be observed on lateral neck roentgenograms. Under ordinary circumstances these may vary in the angle of inclination from about 30 to 60 degrees, but as a rule this does not contribute very much in the way of clinical information. Their delineation becomes of considerable importance in the demonstration of congenital malformations of the base of the skull and the upper cervical segments.

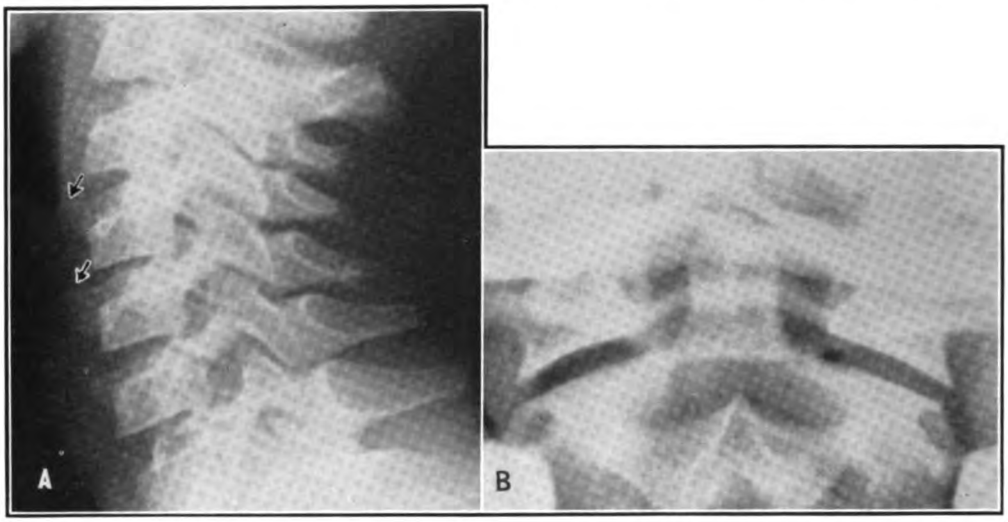


FIG. 27.—*A*, Normal cervical spine in twelve-year-old child. Note shadows of transverse processes projecting in front of vertebral bodies (arrows). *B*, Same patient, anteroposterior view of atlas and axis. Thin white streak in midportion of dens represents line of ossification.

The relationships between the articular surfaces of the first and second cervical vertebræ is of interest. In the normal individual these edges are in correct anatomic position when they appear to be in the same line. However, if the head is turned forcibly to the right or left side, or if there is a pronounced spasm of the occipito-cervical muscular groups, it is not uncommon for the edge of one of the articular surfaces of the first cervical vertebra to be shifted laterally for 2 to 3 mm., while its opposite fellow comes to overlie the articular surface beneath by the same distance. Caution should be exercised in interpreting such variations from the normal as dislocations. As seen in the lateral projection, the tip of the odontoid process may rest at the level of the superior portion of the anterior arch of the first vertebral body, or may be slightly above or beneath it, and space between the articular surfaces may be as much as 2 to 3 mm. The relationship between the tip of the odontoid process and the base of the skull is of importance in the diagnosis of basilar impression. A line drawn between the dorsal aspect of the hard palate and the posterior rim of the foramen magnum (Chamberlain's line) passes above the odontoid tip under normal conditions. If the odontoid extends above this line some degree of basilar invagination presumably is present. Occasionally a variable radiolucent notch-like shadow appears in the medial portion of the articular mass of the atlas, caused by the margin of the medial tubercle of the lateral mass continuous inferiorly with the articular surface of the inferior articular facet and laterally by the concavity of the superior facet. This is attributed in part to the nutrient artery entering the atlas just posterior to the medial tubercle (Meghrouni and Jacobson, 1959). Another variable structure of anatomic interest is the presence of an anomalous ossification center in the oblique atlanto-occipital ligaments. When this calcifies a bony arch is formed which encloses the posterior atlantoid foramen and through which the vertebral artery and the suboccipital nerve passes. Pyo and Lowman (1959) found this to be present in 12.6 per cent of normal skulls. In 4 per cent of their series of 300 skulls this structure was unilateral. In some instances complete arching was present while in others the fusion was incomplete. A wide range of anatomic variation in the "ponticulus posticus" was found to be existent.

As seen in the lateral projection, the antero-inferior aspect of the second cervical vertebra frequently overlaps the corresponding superior surface of the body of the third cervical vertebra. Similar change of lesser degree may be observed between the third and fourth, fourth and fifth and sometimes between the fifth and the sixth cervical segments. The tips of the transverse processes of the cervical segments also overlap the anterior aspects of the bodies of the cervical vertebræ and the respective intervertebral discs. These overlapping shadows may be distinguished on oblique as well as the lateral roentgenograms of the cervical spine. The foramen transversarium of the second cervical vertebral transverse process overlying the anterior aspect of the inferior margin of the second cervical vertebræ is frequently visible and the corresponding foramina of the transverse processes of the third, fourth and fifth cervical vertebræ may also be demonstrable in the lateral view. The intervertebral foramina are best observed on the oblique roentgenograms, preferably right and left anterior oblique stereoroentgenographic studies. On these the foramina on the side near the film are visualized. Rarely, one may observe an additional unfused epiphysis on the inferior lamina of the lowermost cervical vertebræ, or a pedicle may reveal a thin cartilaginous line. These should not be confused with fractures.

The relative positions of the cervical vertebral bodies in flexion and extension may vary considerably in different individuals, depending largely upon the body habitus and muscular propensities of the individual. Thus, a patient with a long slender neck may show a much greater degree of flexion and extension than a short stocky individual with a short neck.

During flexion and extension the articular facets of the cervical vertebræ glide forwards and backwards, resulting in the normal individual in relatively smooth spinal curves. The range of sliding of the various cervical facets is progressively more pronounced in the articulations between the second and third, the third and fourth, the fourth and fifth and sometimes in the fifth and sixth cervical neural arches. In viewing flexion and extension studies of the cervical spine it should be kept in mind that these motions produce relatively parallel change in the alignment of the respective articulating surfaces with a slight forward position of the upper segments. Even small changes in these articular spaces under various types of stresses and strains may be of clinical importance in evaluating the possibilities of injuries with consequent subluxations.

In viewing studies of the cervical intervertebral foramina, one must keep in mind the position of the head during the exposure. With the patient at rest and the head in the neutral position, a properly exposed and positioned film will show each intervertebral foramen from the second down to the upper thoracic region as symmetric, smooth, somewhat oval shadows. Films made with the head bent forward and either elevated or depressed may cause variations in the configuration of the intervertebral foramina in the middle and lower cervical regions. Such distortions should not be considered of a pathologic nature. In addition to this, one not unfrequently observes a rather prominent shadow corresponding to the articular facet between the fourth and fifth, fifth and sixth and occasionally between the third and fourth cervical vertebræ. These appear to impinge into the dorsal aspect of the respective intervertebral foramina. However, studies made on the same patient in the proper position show that these shadows are variable, and they should not be given pathologic significance.

In the lateral projections, the position of the spinous processes sometimes form an indication as to restriction or unusual degrees of mobility of the cer,vical spine. Under normal circumstances, the tips of the spinous processes of the middle group of cervical vertebræ are practically in opposition to each other with the head in extension. In flexion a definite separation of the various spinous processes occurs. Their tips present characteristic configurations, that of the second being bulbous and prominent, the third, fourth and fifth being shorter and smaller, the sixth larger in size, while the seventh is the most prominent of the group. The tips of the spinous processes not infrequently are bifid and irregular. On routine roentgenograms of the cervical spine, these structures may be obliterated by overpenetration because of their relative thinness. However, this can be minimized by viewing the films through a bright light, or if structural changes are suspected, underexposed roentgenograms may prove helpful. They may be identified on anteroposterior films as tear-shaped shadows with calcific rims lying in the midline at a point beneath the inferior margin of the vertebra of origin. The transverse processes of the first cervical vertebra are blunt and heavy, and are pierced by the foramen transversarium. The transverse process of the second cervical vertebra usually is considerably smaller and anterior in its position, so that on oblique roentgenograms it may lie in front of the body of the second and third cervical vertebræ with the foramen transversarium overlapping or in front of the anterior aspect of the second cervical vertebra. Overlying the posterosuperior aspects of the vertebral bodies may be superimposed the shadows of the pedicles. The shadows of the tips of the transverse processes may appear somewhat in front of the respective anterior margins of the vertebræ because of their foward inclination.

Flexion and extension studies of the cervical spine are essential in studying the gliding motions of the vertebral bodies upon each other as well as the changes in the relative position of the articular facets. The range of motion is variable, being greatest in thin individuals with long necks. There is a slight forward motion of the inferior aspects of the second to sixth cervical segments in extreme flexion, with a corresponding shift of the posterior margins. With extension these surfaces glide posteriorly. The intervertebral discs may show slight narrowing anteriorly and posteriorly in flexion and extension respectively. Such films should be preceded by lateral studies in the neutral positions to see if the normal lordotic cervical curve is retained. The articular facets of the cervical spine present the greatest range of motion in the midcervical area.

It has been estimated that the cervical nerves occupy from 20 to 25 per cent of the available area of their respective intervertebral foramina. The remainder of the space is occupied by fatty tissues, lymphatics and blood vessels. With the neck in the neutral or flexed position, the intervertebral foramens are fairly large, but in extension the foramina decrease in size as the gliding surfaces of the articular facets pass over each other as inclined planes. This is most pronounced in the midcervical region where the range of motion is greatest.

THE THORACIC SPINE

Examination of the thoracic spine during the infancy is rarely required except for the identification of congenital abnormalities or the effects of

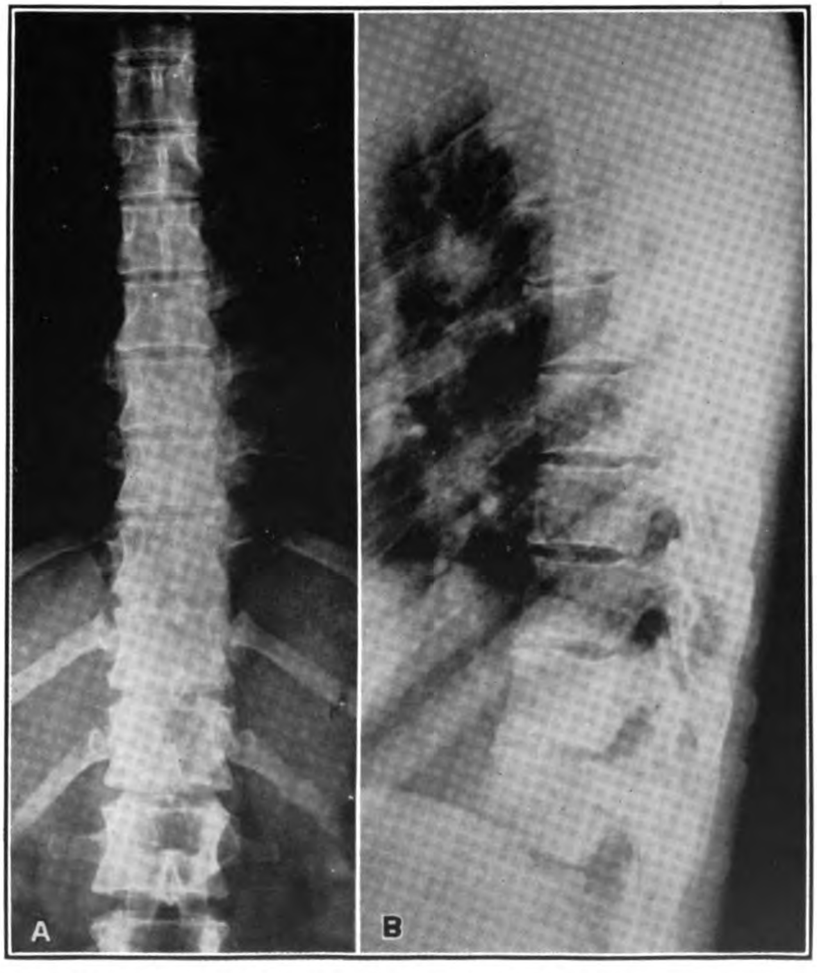


FIG. 28.—*A*, Anteroposterior; *B*, lateral views, normal adult thoracic spine.

trauma. Parenthetically, it might be stated that the dorsal spine in infants is usually visible to a great extent on lateral roentgenograms of the chest, and should be scrutinized as part of such examinations. The demonstration of incomplete fusion of the spinous processes of the upper dorsal segments is not at all uncommon and is of no pathologic importance.

In children and adults, examination of the dorsal spine is usually accomplished by anteroposterior and lateral roentgenograms. Steroscopic views are advised whenever detail as to configuration of the pedicles or the neural arches is desired. In addition, it is sometimes found advantageous to obtain oblique roentgenograms of the dorsal spine when information concerning the costovertebral articulations or the articular facets is required. In obtaining the anteroposterior views of the thoracic spine, the influence of the difference in the anteroposterior diameter of the lower half of the chest compared with the upper half must be taken into account. It is sometimes advisable to obtain studies of the upper half and lower half of the dorsal spine on separate films and the use of cone-down roentgenograms for the sharpest possible detail is advocated. Lateral roentgenograms of the

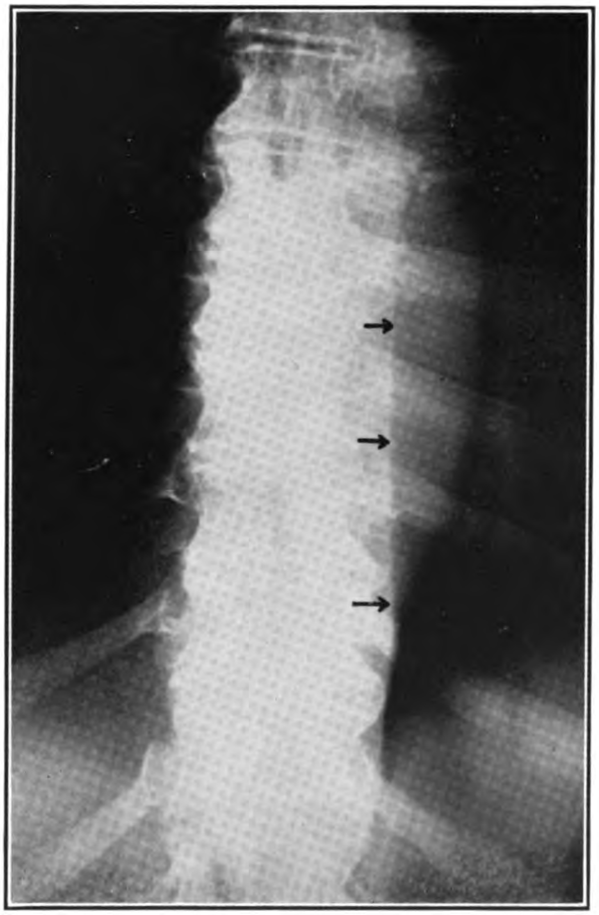


FIG. 29.—Anteroposteror view, normal thoracic spine. Left paravertebral pleural reflection line (arrows) lies medial to arcuate shadow of descending thoracic aorta.

dorsal spine will usually visualize the third to the tenth dorsal vertebræ very adequately. Special technics, however, are required for the proper demonstration of the first, second and third thoracic vertebræ. In such studies a modified oblique projection is utilized, the lower cervical and upper dorsal segments being seen through the thoracic inlet. I have found that laminagrams of the upper dorsal vertebræ in the lateral view sometimes portray these structures to better advantage than the routine technics. In the evaluation of the lower dorsal segments, one must consider the relative position of the diaphragm and the abdominal viscera. If these overlap the lower dorsal vertebræ, a technic which shows the middle thoracic vertebræ will be too light, and additional views coning down over the lower dorsal segments are required.

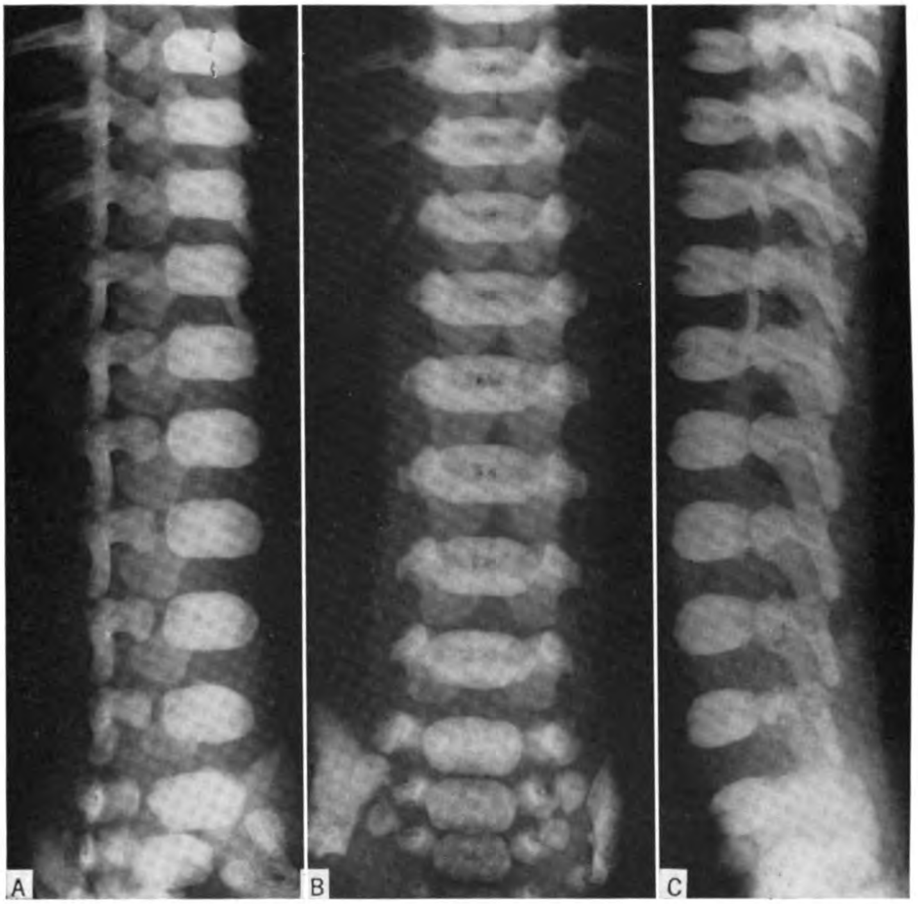


FIG. 30.—*A*, Oblique roentgenogram of lower thoracic and lumbar spine, excised specimen. The neurocentral synchondroses are ununited.

B, Anteroposterior roentgenogram, same specimen. The laminæ are visible, but the spinous processes are not calcified. The radiolucencies in the center of the respective vertebral bodies are due to vascular channels.

C, Same specimen, lateral view. Note the anterior notches in the midportion of the vertebral bodies. These are due to venous reservoirs within the spongiosa, which later disappear. Indentations into the posterior aspect of the vertebral bodies at the same level mark the entrance of the posterior vertebral veins and nutrient arteries, and persist into adult life.

It is not necessary to repeat the anatomical details which render the dorsal vertebræ different from the remainder of the spinal column. It should be recalled, however, that the spinal canal in the dorsal area is circular in configuration and smaller than seen either in the cervical or lumbar regions. The marked thickness of the neural arches of the dorsal spine together with the overlapping of these structures should also be recalled. The spinous processes of the dorsal spine are also different from those of the remainder of the vertebral column and the difference in their configurations may serve to identify the respective thoracic vertebræ.

The intervertebral foramina of the dorsal spine are relatively smaller than either the cervical or lumbar intervertebral foramina. Relatively little adipose tissue is contained in the intervertebral foramina of the dorsal spine, since little protection of the nerve root is required because of the relative lack of motion in these joints.

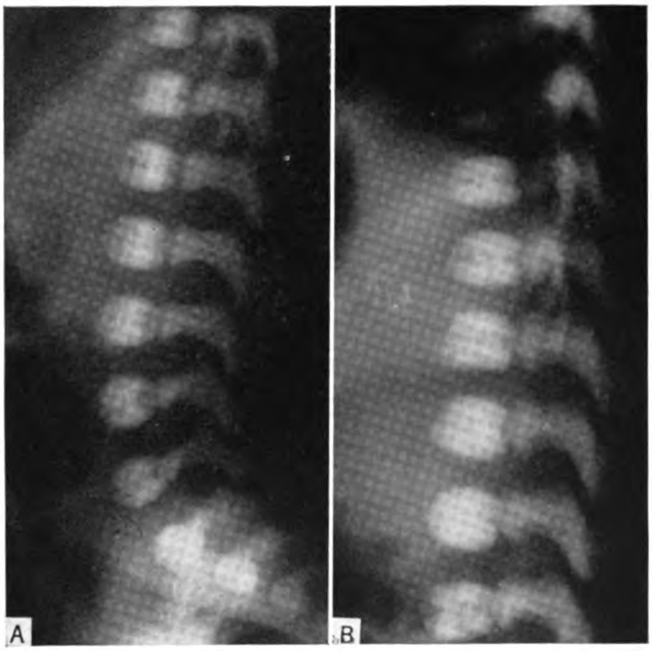


FIG. 31.—*A* and *B*, Lumbar vertebræ in normal newborn infants. Note the variability in the contours of the bodies, the open neurocentral synchondroses, the vascular grooves, and the width of the spinal canal.

On examining the roentgenograms of infants and sometimes children, one not infrequently observes the presence of notches in the mid-portions of the bodies of the lower dorsal vertebræ. The anterior notches usually disappear by the first year of life, but the posterior notches may persist even into adult life. These markings are due to the presence of sinusoidal venous reservoirs located within the spongiosa of the vertebral body, and markings are not of clinical importance.

Beginning at approximately the sixth year of life a ring apophysis appears on the superior and inferior surfaces of the middle and lower thoracic vertebræ and the upper three or four lumbar vertebræ. Occasionally such apophyses also appear in the lower cervical and upper thoracic regions.

These are represented as incomplete thin rims of bone slightly apart from the vertebræ, visible both in the lateral and anteroposterior views. Ossification of these structures is most pronounced at about the age of thirteen years, and fusion with the vertebral bodies occurs by the time the patient reaches the age of eighteen. These structures lie outside the true vertebral epiphyseal plates and maintain their own rate of growth, unassociated with the growth of the vertebral body itself. (Fig. 16, p. 39).

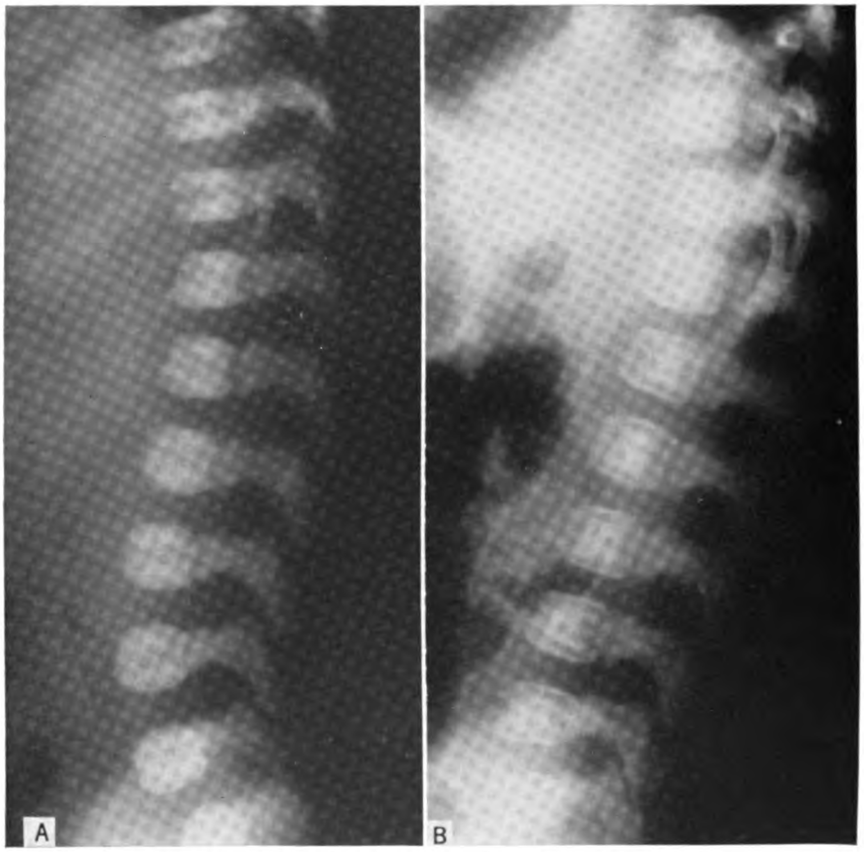


FIG. 32.—*A*, Lateral roentgenogram of normal thoracolumbar spine of one month old infant. Calcification heaviest at lower and upper aspects of vertebral bodies. *B*, Lateral roentgenogram of normal thoracolumbar spine of 8 weeks old infant. Calcification heaviest at centrum. Normal variant.

Inspection of the paravertebral regions on anteroposterior roentgenograms of the dorsal spine is often of considerable clinical importance. To the left of the thoracic vertebræ one can quite frequently observe a line of increased density. This is more often present on the left than on the right side, and may extend from the level of the fourth thoracic vertebra to approximately the level of tenth or eleventh dorsal vertebra. It has been established that this line represents the reflection of the medial pleura of the left lung when seen on the left side. In this situation it should be differentiated from the margin of the descending thoracic aorta which usually lies laterally and presents a more oblique course in the chest, being situated more laterally in the uppermost aspect and medially in the lowermost aspect as it comes to approach the region of the aortic hiatus of the diaphragm. In the lower right thorax, because of shelving characteristics of the posteromedial aspect of the lung due to the presence of the azygos vein, the right posteromedial pleural line is usually not as apparent, and may appear to be continuous with the shadow of the psoas muscle below the right crus of the diaphragm. (Lachman, 1942.)

Attention is again called to the necessity of scrutinizing carefully each pair of pedicles in the dorsal spine as well as in the other areas of the vertebral column. Widening of the interpedicular spaces as measured between the innermost portions of the respective pedicles may provide significant diagnostic information. In view of the relative frequency of spinal cord tumors in the thoracic area, these structures assume an importance which should be clearly recognized.

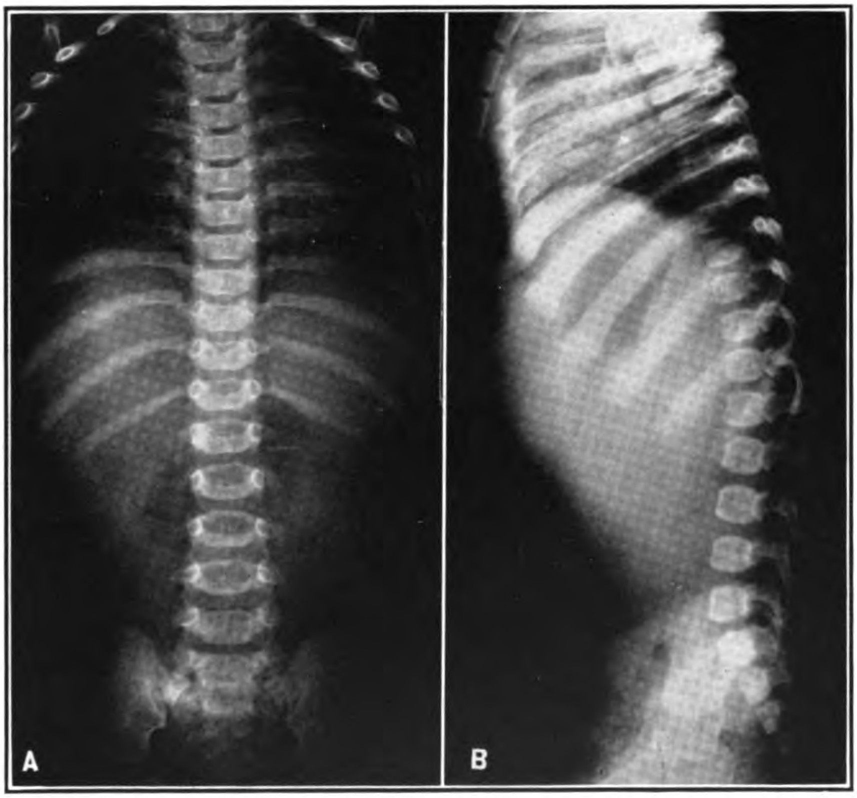


FIG. 33.—*A*, Anteroposterior; *B*, lateral views of normal spine, six-month-old infant. Note increased density of peripheries of vertebral bodies and residual small central area of increased calcification.

THE LUMBOSACRAL SPINE AND COCCYX

Radiographic examination of the lumbosacral spine and pelvis in the newborn is largely accomplished with anteroposterior and lateral roentgenograms. Oblique roentgenograms may be taken if there is any question as to the configurations of the neurocentral synchondroses. During the newborn period, the bodies of the lumbar vertebræ present a biconvex

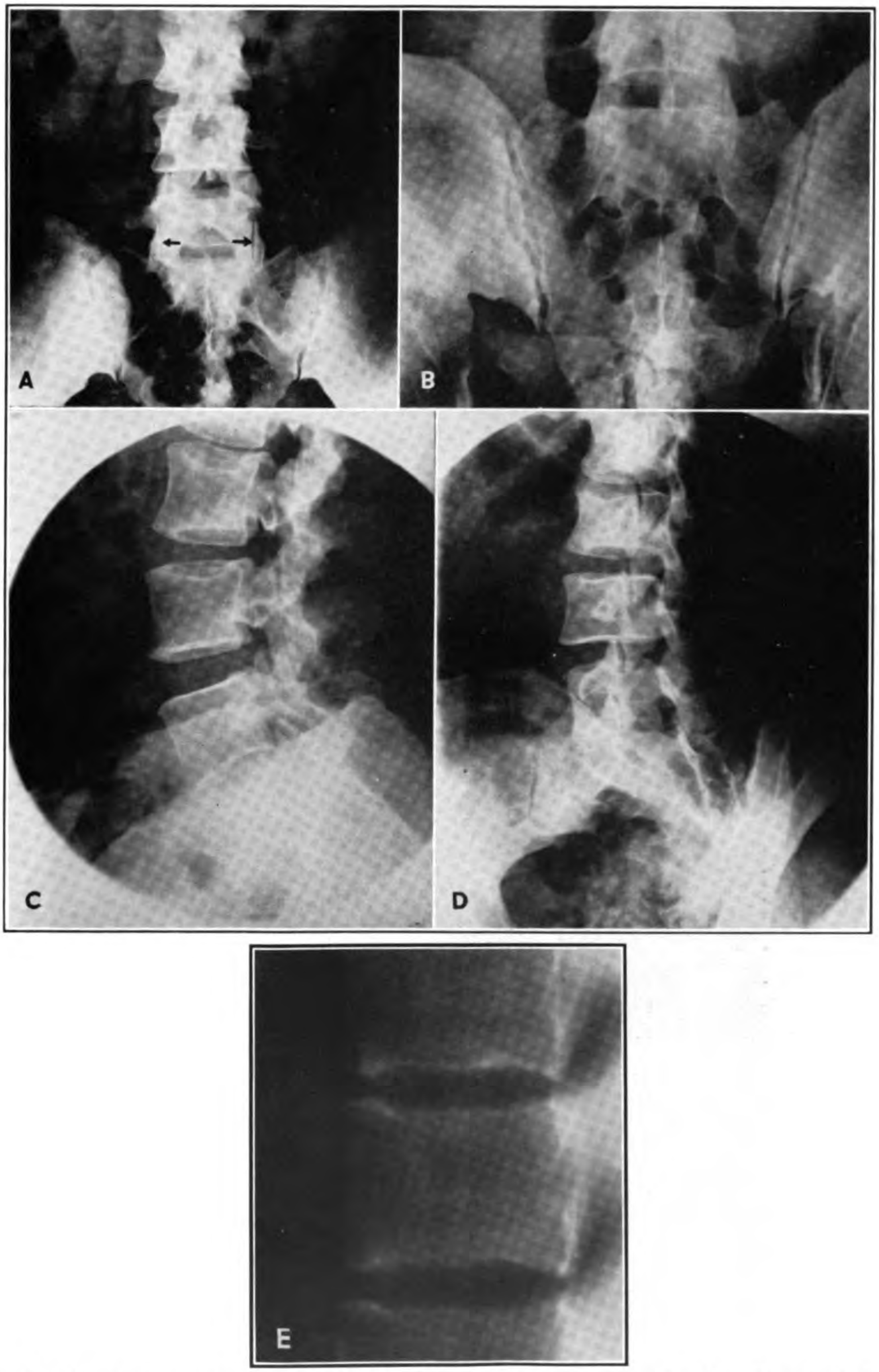


Fig. 34.—*A*, Anteroposterior view, lumbosacral articulation, normal adult spine. Note vertical position of articular facets (arrows). *B*, Same case, 45 degree tilt-up view. *C*, Same case, lateral conedown view of lumbosacral articulation. *D*, Same case, posterior oblique view showing articular facets. The right posterior oblique projection brings out the left articular facets. *E*, Body section roentgenogram of the body of the ninth thoracic vertebra of a 37 year old man demonstrating the posterior notch for the passage of the basivertebral vein.

appearance with a relatively straight anterior and posterior wall. The appearance of the vertebral bodies varies considerably in normal infants. In some instances the remnants of a center of ossification may be seen in the middle of the spongiosa of the vertebral bodies while their peripheries are

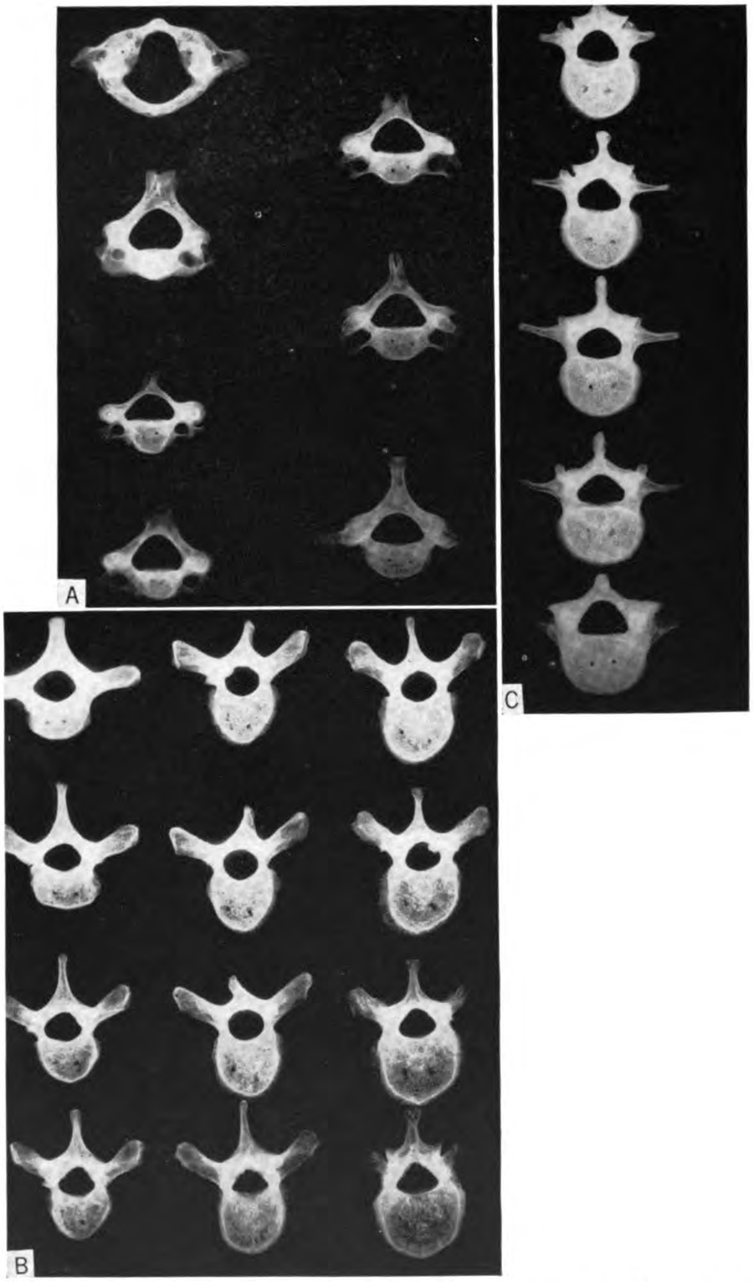


FIG. 35.—*A*, Normal young male adult cervical spine demonstrating spinal foramina. C-1 upper left, C-7 lower right. *B*, Same spine, thoracic vertebræ. T-1 upper left, T-12 lower right. *C*, Same spine, lumbar vertebræ. L-1 top, L-5 bottom. *D* and *E*, Comparison of normal adolescent and elderly spondylotic lowermost three lumbar vertebræ. The latter has a narrowed canal. Note the constriction at the lateral angles of the spinal foramen at L-5 in the adult vertebra.

thin and sharply defined. In others the vertebral bodies present plates of dense bone in their superior and inferior aspects, separated by less densely calcified spongiosum. Fissures are seen at the middle of the anterior and posterior aspects of the middle and lower thoracic vertebræ and the upper

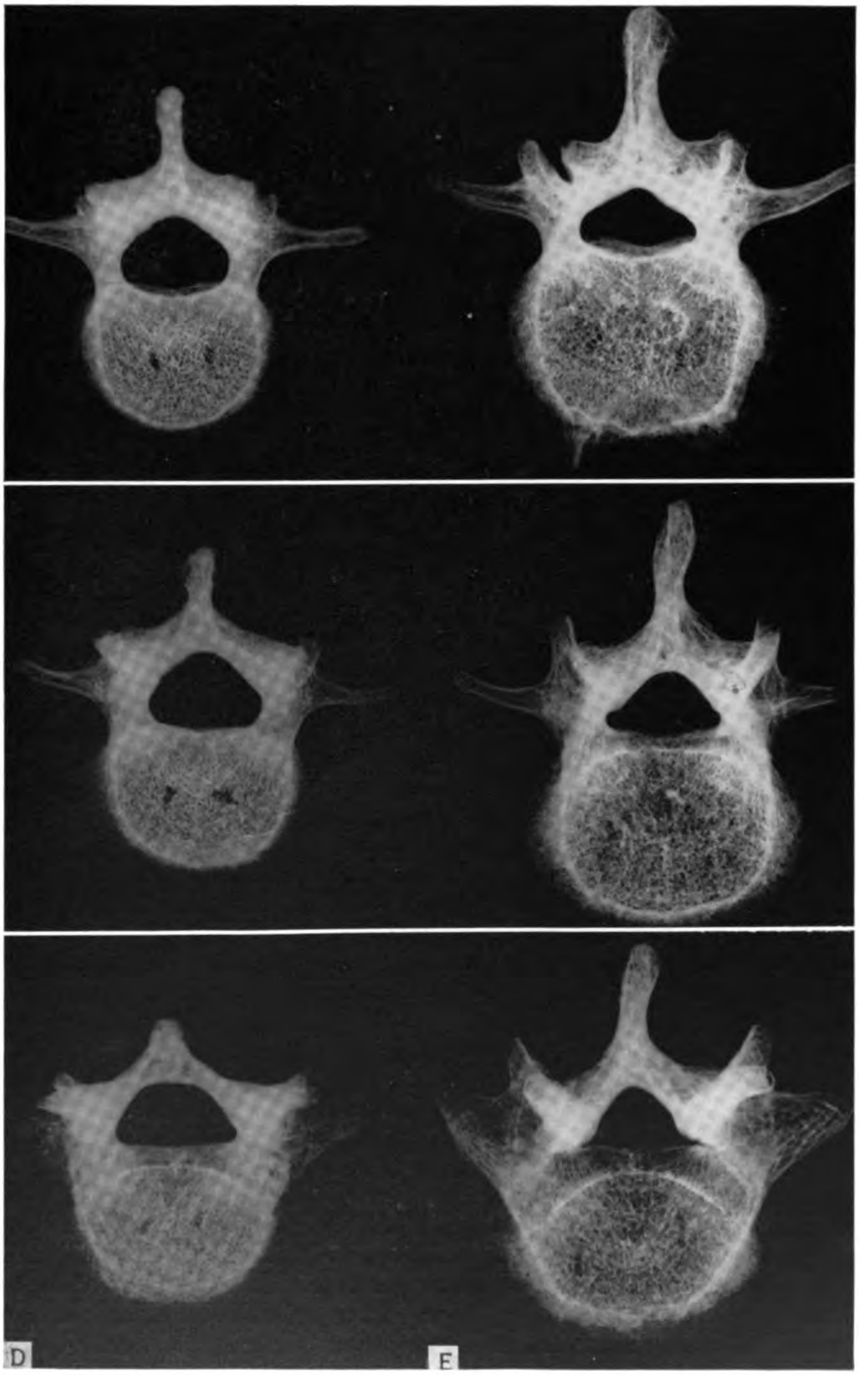


FIG. 35.—(*Continued*).

lumbar vertebral bodies corresponding to sinusoidal venous reservoirs. The neurocentral arches are incompletely fused, the pedicles are relatively heavy and the laminæ are well formed but are poorly seen because they are still largely cartilaginous. The spinous processes, being unossified, cannot be well demonstrated.

On the lateral views there is usually a slight lumbar curve convex anteriorly at the lumbosacral junction. The intervertebral discs are relatively large and are wider at their anterior and posterior aspects. The intervertebral foramina likewise are quite large. The spinous processes are not visible.

The intervertebral discs increase in width slightly from the upper to the lower lumbar areas. At no time have I seen a narrowing of the lumbosacral interspace in the newborn period. The bodies of the sacral vertebræ are separated from each other by intervertebral discs which are better identified on lateral than on frontal radiographic examinations. In the newborn period the transverse processes may be unfused with the vertebral bodies, and are paired off in relationship to each of the central five structures. The first segment of the coccyx may be ossified at birth, but the remaining coccygeal components usually are not visible in the newborn. The dorsal plate of the sacrum, corresponding to the neural arches, can be identified on lateral films, and likewise are separated by radiolucent cartilaginous bands.

During childhood the ossification of the lumbar vertebræ proceeds slowly. By approximately the third to fourth year of life, the neurocentral synchondroses fuse with the vertebral bodies. The ossification of the neural arch proceeds so that articular facets and laminæ become increasingly visible because of the increased calcium content. The spinous processes likewise begin to appear radiologically and are quite well seen by the time the fifth or sixth year has been reached. The anterior sinusoidal venous channels gradually vanish, while the posterior counterparts may remain visible well into adult life.

Radiologic examination of the lumbar spine from childhood to old age may be accomplished in a variety of ways, each designed to provide information concerning some specific point. A routine roentgenographic examination of the lumbosacral spine should consist of anteroposterior and lateral views, and when indicated oblique views for demonstration of the articular facets and the neural arches. If information concerning the mobility of the spine is required, studies may be made in the erect frontal views with the patient bending sharply towards the right and left side for demonstration of change in the interspaces between the various lumbar vertebræ. Additional lateral studies should be made in flexion and extension. A variety of views have been proposed for examination of the lumbosacral articulation, sacrum and sacro-iliac joints. The author believes that this is best accomplished by means of cone-down roentgenograms in the lateral and oblique projection, centering on the lumbosacral articulation or midsacrum as required and with so-called "tilt up" views of the sacrum and lumbosacral joints in the anteroposterior position. The latter have to be made with consideration of the configuration of the sacrum in the particular individual. As a rule, it is possible to obtain a study of the sacrum in which the intervertebral foramins are rendered as circular or almost circular shadows if the tube is tilted approximately 15 to 25 degrees towards the patient's head with the central ray passing through the body of the first sacral vertebra. Stereoscopic views of the lumbosacral spine and the sacrum have been found helpful in identifying areas of altered bony density. Laminagraphic studies are also quite useful, particularly in demonstrating small areas of bone destruction. Their application to studies of the lumbosacral joint and sacrum has been found to be quite satisfactory in lateral views which portray the dorsal and ventral walls of the sacral canal to better advantage than can usually be observed even on the best straight films. The sacro-iliac joints are best seen on the tilt-up views, but oblique views have also been recommended. Anteroposterior laminagrams of the sacrum and of the sacro-iliac joints may prove informative. The configuration of the spinal foramen of each lumbar vertebra has definite clinical importance. The first and second

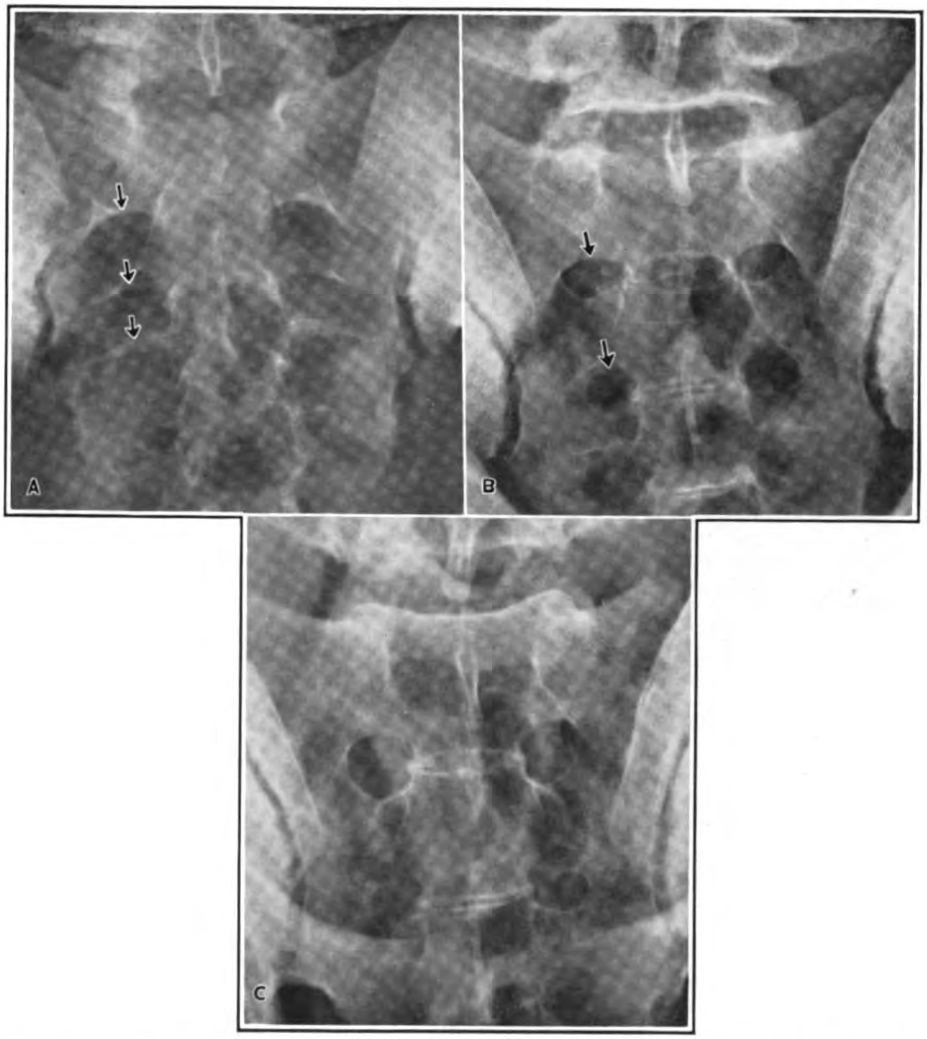


FIG. 36.—*A*, Direct anteroposterior view of normal adult lumbosacral junction. The sacral foramina are foreshortened, and their superior rims are presented as rather dense lines (arrows). *B*, Same patient, tiltup view with beam directed thirty degrees towards patient's head. The lumbosacral joint is better visualized, and the sacral foramina are projected as oval structures (arrows). *C*, Same patient, 45 degree tiltup view. The upper two sacral foramina are practically circular. Note the difference in the appearance of the sacroiliac joints on the three projections.

lumbar vertebral canals are rather triangular, with the lateral aspects well rounded. The last three lumbar vertebral foramina tend more to a pentagonal configuration, but their heights are less than the upper two. Measurements made on lateral roentgenograms indicate that the average height of the first two lumbar spinal foramina is approximately 14 to 22 mm., while the lower three is between 13 and 20 mm. From the clinical viewpoint the dimensions of the lateral aspects of the spinal canal are most significant because it is here that diminished space results in nerve root compressions. Unfortunately, these areas cannot be measured, but some inference as to the available space sometimes can be made by determining the anteroposterior measurements. If significantly narrowed, particularly in the lowermost two lumbar segments, where we regard a measurement of less than 14 mm. as significant, the effects of spondylotic ridges or herniated discs may be augmented.

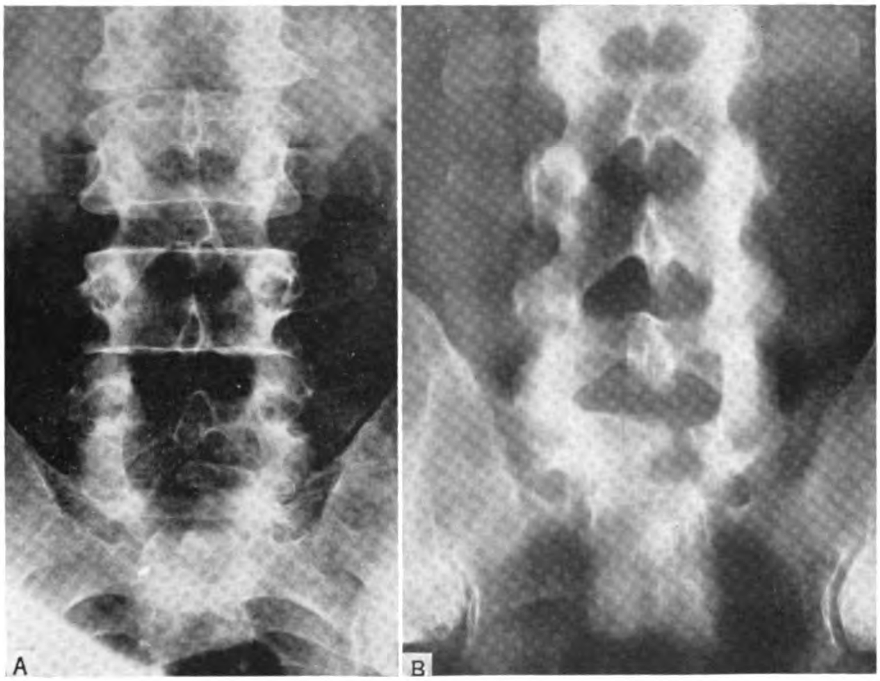


FIG. 37.—*A*, Anteroposterior roentgenogram of lumbar spine. Incomplete fusion of spinous process of L-5. *B*, Same patient, body section roentgenogram. The neural arches are sharply in focus, and the malformation of the spinous process of L-5 is better seen.

A study of the lumbosacral articulation alone could well be the subject of a monograph. The multitudinous variations discovered as incidental findings during roentgenographic examinations made for purposes other than study of the lumbosacral spine seem to indicate that many anatomic variations may exist in this area without clinical evidence of impairment of spine function. However, well-qualified observers have come to other conclusions as to the significance of minor lumbosacral articulation malformations. Howard (1942) mentioned that symmetrical articulations of the lumbosacral apophyseal joints may predispose to back pain. Ferguson (1934) stated that sagittal positions of the articular facets of the fourth and fifth lumbar vertebræ and fifth lumbar and first sacral vertebræ were advantageous from a mechanical point of view, while an anteroposterior position (coronal articular facets) was a predisposing factor to weakness of the structural function of the lower lumbar spine and might be a causative factor in the production of pain. He was of the opinion that mixed or asymmetrical articular facets in the lumbosacral spine predispose many people to pain because of weakness of structure. The incidence of pain was found to be more frequent in people with narrow lumbosacral interspaces in a ratio of 4.5 to 1 by Hodges and Peck (1937). In individuals with low back pain developmental anomalies in the lumbosacral region occurred with an increased frequency of 2 to 1. On the other hand, Mitchell (1934) mentioned that asymmetry of articular facets of the lumbosacral spine was favorable in that it permitted a certain degree of rotation, whereas purely sagittal or coronal arrangements prevented such movement. It was his opinion that arrangement of articular facets in a coronal plane caused these to impinge upon one another so that any tendency towards forward slip was inhibited.

Pheasant and Swenson (1942) concluded that the lumbosacral articulations in normal individuals were arranged most frequently in an oblique or coronal place. Purely oblique or sagittal planes of articulation were not encountered. Roentgenologically the facets may appear to be situated coronally or sagittally depending on which component predominates. They concluded that asymmetry of the lumbosacral region occurs in approximately one-twenty-fifth of all human spines, varying in degree and accompanied by alterations in force transmission of varying significance.

It is the author's observation that narrowing of the lumbosacral interspace in itself is not of any clear-cut diagnostic significance. It has also been my experience that the demonstration of articular facets of the lumbosacral junction in an oblique or sagittal plane does not necessarily indicate the cause of the patient's complaints. A review of a long series of spine studies in patients admitted for conditions other than back pain, as for instance for intravenous urography or gastro-intestinal complaints, showed that many normal individuals present minor congenital variations at the lumbosacral articulation. I do not, therefore, give great significance to the demonstration of a narrowed lumbosacral interspace or to variation in articular facets between the fourth and fifth lumbar and fifth lumbar and first sacral segments as a definite cause for pain. In those instances the cause for pain must be sought for in a disturbance which may not be visible on plain roentgenograms. The first thing that comes to mind in such circumstances is the possibility of torsional and stress disturbances of the soft tissues of the lower lumbar and lumbosacral articulations. The possibility of disease of the intervertebral disc especially discal herniation must be considered with recurrent pain, especially if radicular symptoms are present.

Congenital malformations of the neural arches of the first, second or third lumbar vertebræ are infrequent. An exception is the demonstration of small fissures across various portions of the arch or the articular facets, usually across the distal portion of the inferior articular facet. In 1939, Hipps had the opportunity to examine such an anomaly at operation. Microscopic and gross studies of this anomalous bone showed that the zone of rarefaction was not a fracture, but a developmental abnormality in the nature of an apophyseal line. This fissure line was found to be composed of hyaline cartilage, and the microscopic picture of this region ruled out the possibility of an ununited fracture, or so-called "Umbauzon."

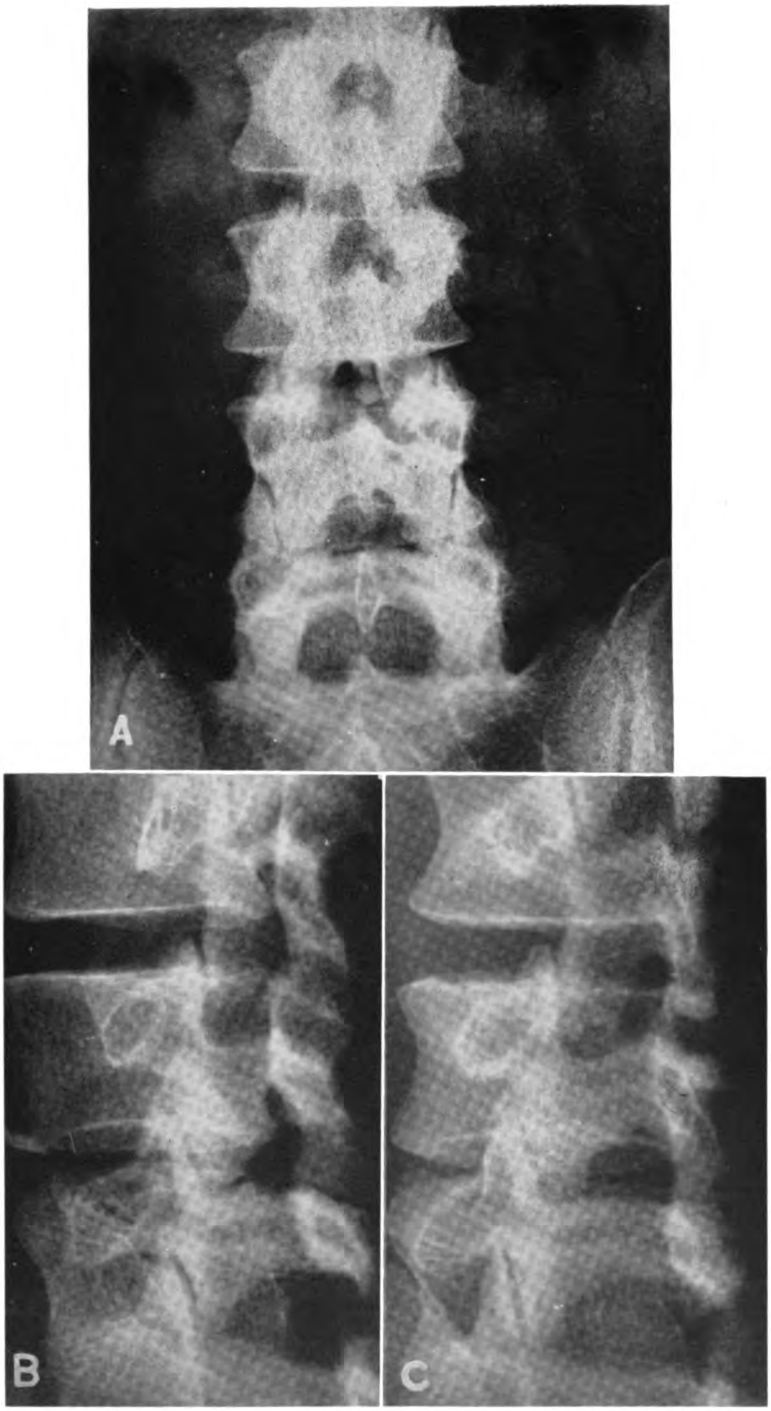


FIG. 38.—*A*, Anteroposterior view, normal adult lower lumbar spine. The articular facets of the two presacral vertebræ are in different planes. The lumbosacral facets are coronally placed, and those between the fourth and fifth lumbar vertebræ are sagittally situated. Both are normal. *B* and *C*, Right and left anterior oblique views. The slight haziness of the articular facets between the fourth and fifth lumbar vertebræ is not pathologic. Note the clarity of the facets between the fifth lumbar and first sacral vertebræ. An accessory epiphysis is present at the inferior aspect of the inferior articular process of the third lumbar vertebra on the left side.

According to Hipps these fissures represented failures of fusion of one of the normally present ossification centers lying at the base of an articular facet. In a review of the literature, he found that the lesion was usually unilateral, but occasionally bilateral, and was more often observed on the right than on the left side. The superior articular facet was rarely involved and most cases were seen in the second and third lumbar neural arches. Males were more frequently affected than females. His figures were approximately the same as reported by Farmer in 1936.

Oppenheimer (1941, 1942) also studied fissure formation in the vertebral articular processes and pointed out that they may occur in the laminæ, in the isthmus between the superior and inferior articular processes, as well as at the tips of the inferior articular processes of the lumbar vertebræ. In general each articular process has one ossification center for its main part, and sometimes a second one for its tip. In most instances the articular segments which consist of the superior articular process, the isthmus and the inferior articular process, have two constant and two inconstant centers. If the bones arising from a center fails to unite, the cartilaginous layer between may be seen roentgenologically as a radiolucent streak. Rarely, additional centers of ossification may exist. A longitudinal line may extend from the isthmus through the articular process dividing the latter into halves, indicating that each articular process may have three ossification centers—two in its main part and one in its tip. Together with the ossicle at the isthmus recorded in this communication by Oppenheimer (1942), the number of separate bones in each articular segment may total seven. This phenomenon should not be confused with fracture lines.

The angle of inclination which the sacrum assumes in relationship to the lumbosacral articulation is of clinical interest. It was pointed out by Ferguson (1949) that the normal angle formed in the horizontal plane with the articular surface of the first sacral segment is 34 degrees as seen on a true lateral film. If this angle is significantly exceeded, it is Ferguson's opinion that instability of the lumbosacral junction exists.

Anomalies of articulation of the fifth lumbar vertebra in relationship to the first sacral segment are frequent. One often observes a transitional fifth lumbar vertebra in which one or both transverse processes as well as the inferior aspect of the body partially fuses with the sacrum, giving rise to the so-called "sacralized fifth lumbar vertebra." The opposite process in which a first sacral segment is significantly separated from the remainder of the sacrum and is referred to as a lumbarized first sacral segment. Indeed it is not at all infrequent to observe individuals who apparently have 6 lumbar vertebræ. In such instances it is preferable to designate the distal lumbar segment as the first presacral vertebra. If it is desired to definitely establish whether or not such a transitional form exists, a count of the entire spine from the first cervical segment down is necessary.

The structure of the transverse processes of the fifth lumbar and first sacral segments often is of considerable interest, particularly if articulations are formed between the tips of the transverse processes of the fifth lumbar vertebræ and the adjacent sacrum and iliac bones. It is generally believed that if this change is bilateral, no weakening of the lumbosacral articulation exists. However, if it should be unilateral, changes in stress may result in painful syndromes and arthritic manifestations may appear. In my own experience such changes have often been observed in patients who had no symptoms whatsoever, and unless examination fails to disclose any other reason for pain in the lumbosacral region, I would hesitate before deciding

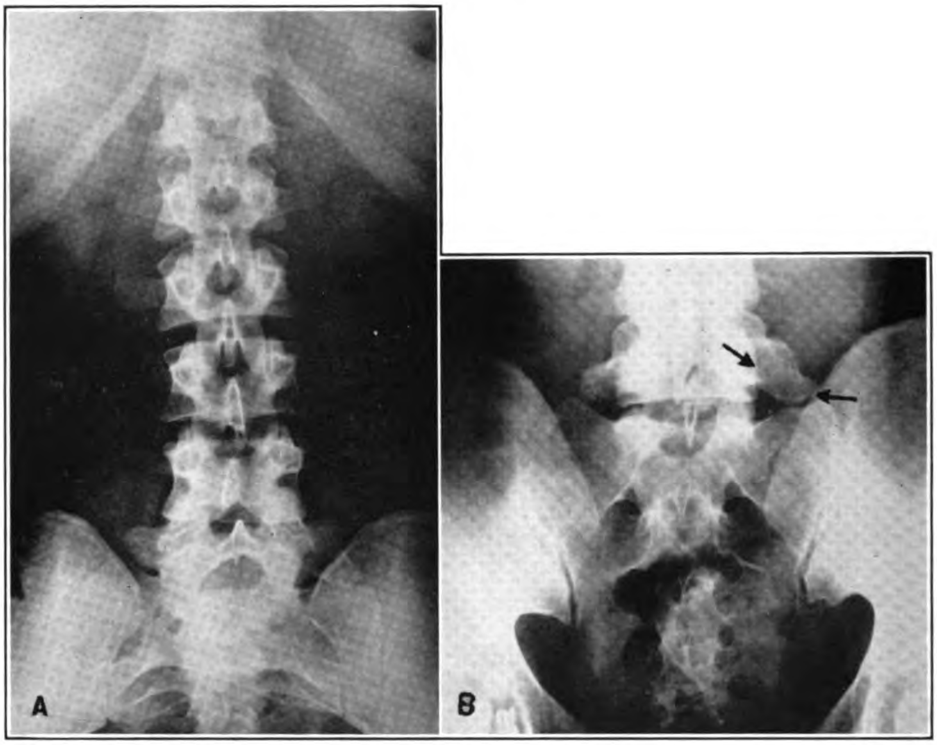
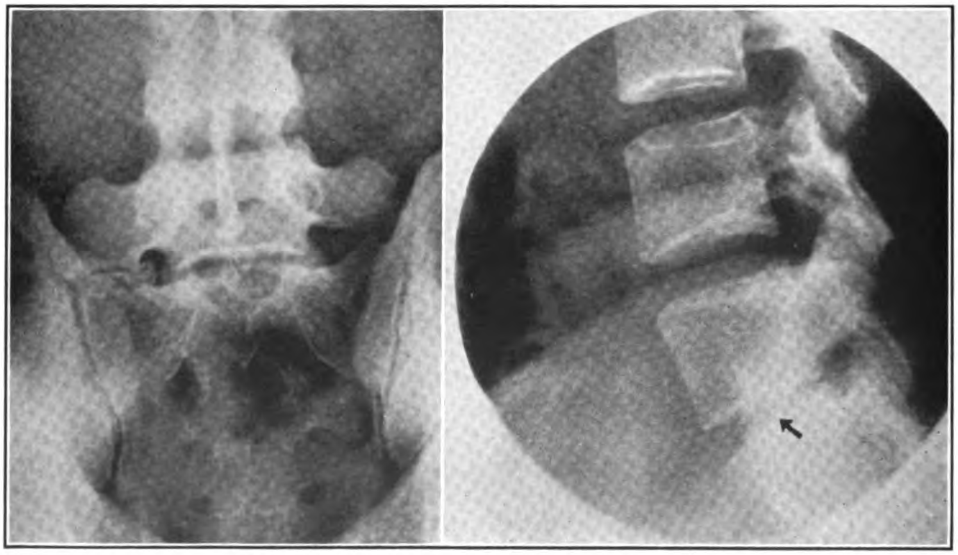


FIG. 39.—*A*, Anteroposterior view normal adult lumbosacral spine. The presacral vertebra is not united to the sacrum, and represents a sixth lumbar vertebra. *B*, Same case, tiltup view. Note the prolonged transverse process of the presacral vertebra (arrows), and the spina bifida occulta of the uppermost sacral vertebra.



A *B*

FIG. 40.—*A*, The right transverse process of the fifth lumbar vertebra articulates with the iliac bone and the sacrum. *B*, Same case, lateral conedown view. The enlarged right transverse process is projected across the lumbosacral articulation (arrow).

that the existence of such an anatomic variation is the definitive cause of the patient's complaints. Calcification of the ileo-lumbar ligaments occasionally appear, and are of no specific importance.

There has been much discussion about the demonstration and importance of abnormal mobility of the lower lumbar spine as a cause of persistent low midback pain. Various patterns of minor shifts of the lower lumbar vertebræ have been reported, based on examinations made with the patient in strained flexed and extended positions. While of interest in demonstrating the wide range of motion, it is noteworthy that many patients have long intervals of freedom from pain with no change in their anatomic architecture which can be demonstrated radiologically. At present no definite correlation between such symptoms, often indefinite and of variable intensity, and the bony configuration of the lower vertebral column can be regarded as established with certainty. Discussion of deformities such as spondylolysis and spondylolisthesis is assigned to a later chapter.

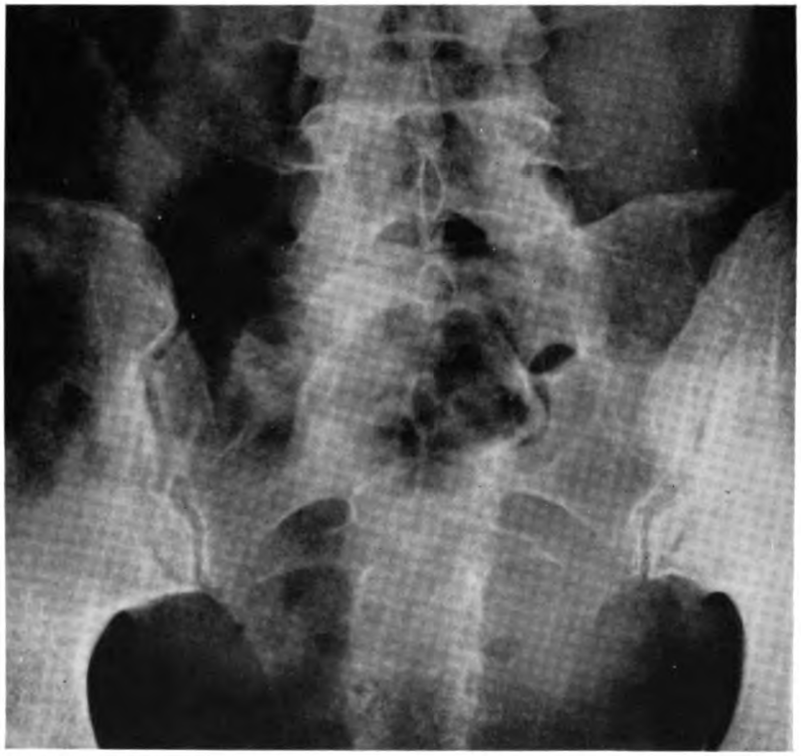


FIG. 41.—Transitional presacral vertebra (sacralized fifth lumbar). Massive left transverse process fused with upper left sacrum and articulating with sacroiliac synchondrosis. Note the calcification in the left iliolumbar ligament.

Variations in the structure of the sacrum sometimes are observed on plain films, and may be of diagnostic importance. Among these are changes in the diameter of the sacral canal pointing to the presence of an occult meningocele, obliteration of the lumen of the sacral canal, shortening or other bony deficiencies of the sacrum or coccygeal abnormalities. Some are of practical interest when caudal anesthesia is contemplated.

The importance attributed to roentgenographic examination of the spine in industry is attested by the reports of pre-employment surveys. O'Connor

(1946) found that pre-employment spine roentgenograms helped to reduce the incidence of back strains by enabling each person to enter a job suited for his own physical capacities. Barton and Biram (1946) reported on routine anteroposterior and lateral roentgenograms of the lumbosacral regions in 1,000 consecutive preplacement examinations. They found 856 defects in 498 patients, and concluded that spondylolisthesis, arthritis and in some cases transitional lumbosacral vertebræ were significant factors in back strain in workers over forty-five years of age. A report on a series of 3,000 preplacement examinations of the lumbosacral spine over a two-year period was made by Allan and Lindem (1950) who encountered a large number of asymptomatic conditions, including degenerative processes and congenital or developmental anomalies. These authors were of the opinion that in industries requiring the use of physical exertion, preplacement examinations of the spine may well be compared with survey roentgenograms of the chest. It would appear reasonable to check any individual who would be required to exert himself or who might be subjected to strains of the back before employment. In this way, incidence of chronic low back pain might be minimized by the avoidance of undue strain in individuals who appear to be susceptible to such disturbances by reason of an anatomic variation in their spines. This statement might appear contradictory in view of what has been said before in the present chapter. In mitigation of this apparent discrepancy, I would remind the reader that the result of a survey of over 1,000 patients is applicable to industry on a broad scale, while an individual's variation has to be evaluated in the light of his existing symptoms.

A few comments may be made here as to the relative importance of the various components of a vertebra in casting a roentgenographic shadow. One must keep constantly in mind the fact that we are dealing with a composite picture, the cortex being relatively radio-opaque, while the spongiosa is considerably more radiotransparent. Snure and Mainer (1937) came to the conclusion that most of the detail of the osseous structures portrayed on roentgenograms are caused by the cortical bone, while the spongiosa accounted for the general density of the vertebral body. They noted that when areas of cortical bone 1 cm. square were removed from the lateral surfaces of a vertebral body and the excavated area was filled with paraffin, their locations could be demonstrated on roentgenograms. Three fine holes extending almost across the vertebral body through the spongiosa, however, when filled with paraffin were practically invisible. On the other hand, Wagoner, Hunt and Pendergrass (1945) came to the conclusion that the spongiosa was the more important component in the production of the roentgenographic shadow pattern of a normal vertebral body. I have on several occasions made roentgenograms of lumbar and thoracic vertebræ in which relatively large holes were present in the spongiosa. Unless these were quite large, exceeding 1.5 cm. in diameter, it was noted that the shadows were obscured by the overlying shadow of the cortex. On the other hand if only a small bit of cortex was removed, the density of the remainder of the cortex as well as the relatively intact spongiosa prevented identification of the injured area. It is difficult to transpose such evidence to clinical terms. I believe that it is fair to state that fairly extensive destruction of either the cortex or the medulla is required to identify the change roentgenologically in the living being. The results of experiments on dead tissue can hardly be transposed because the effect of overlying tissues, water content, motion and strain is eliminated. Thus it is possible for a vertebra to appear quite normal roentgenologically on one examina-

tion, and only a few days later to observe a change in its contour secondary to compression. This may be the first radiologic indication of disease of the bone. The obvious conclusions are that a considerable amount of destruction of either the cortex or the spongiosa must exist before the lesion can be identified *in vivo*, and that roentgenograms of the spine are deficient in demonstrating small diseased areas, even under the best circumstances.

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MYELOGRAPHY

Contrast examination of the spinal canal is accomplished by the introduction of gases which render the subarachnoid space more transparent than the adjacent tissues, or by intrathecal instillation of radioopaque materials. In 1918 air myelography was recognized by Dandy as a method which offered excellent diagnostic advantages. While gas myelography has more or less fallen into disuse, it remains an important method which should be utilized for inspection of the cervical spinal canal routinely with lumbar pneumoencephalography and when the introduction of opaque medium appears inadvisable.

The pioneer work of Sicard and Forestier in 1922 established lipiodol myelography as one of the significant advances in neuroradiology. First conceived of as a therapeutic measure, the originators of this method first instilled lipiodol into the epidural space. Quite by accident they discovered that the material produced no perceptible ill effects when introduced into the subarachnoid space, and that it flowed freely and could be utilized with great accuracy in the diagnosis of spinal cord tumors. The prime objective of the examination was enunciated by them in the statement that "we have been able to guide the hand of the surgeon."

Epidural injection was soon abandoned for the exciting opportunities offered by intrathecal injection of lipiodol, and Sicard, Paraf and Laplane were able to report in 1923 that during the past two years they had explored the meninges of the spinal cord in about 150 patients. They preferred to inject the lipiodol into the cisterna magna as suggested by Ayer because at that time the aim was to assure the fall of lipiodol in the spinal canal, thus permitting identification of various obstructing lesions. However, the advantages of lumbar instillation were also carefully considered.

The possibilities of untoward reactions to the subarachnoid placement of lipiodol came under investigation quickly, and various observers noted that meningeal inflammatory reactions were relatively frequent. Nevertheless, because of the vast importance of the information available from lipiodol myelography clinical investigations continued even though the possibility of inflammatory reactions was recognized. Fluoroscopic observation of the movement of lipiodol within the spinal canal soon became a necessary procedure, and myelography became an acknowledged neuroradiologic method. The superior accuracy of localizing spinal cord tumors over clinical methods was recognized and surgeons began to avail themselves of this information for more precise planning of their procedures.

Thorotrast was suggested by Nosik for myelography, but this technic which included forced drainage for the removal of the thorotrast, never received acceptance. The advantages of absorbable radioopaque materials are strongly advocated by Scandinavian workers. In 1931 Arnell and Lidstrom reported on myelography with Skiodan (abrodil). For a while this was neglected, but about 1944 Arnell and others revived interest in it, so much so that at present it is the method of choice in Scandinavian clinics.

While there are certain technical advantages to the use of water-soluble substances its handicaps include the possibilities of adverse reactions including shock and a few cases with fatal outcome, limitation of the examination to the lumbar region, and the necessity for spinal anesthesia as a preliminary step. Certain authorities are quite enthusiastic about its fluid qualities in portraying nerve roots. The examination is made only by films and this must be done within fifteen minutes before the contrast agent is absorbed.

With recognition of the importance of herniation of the intervertebral discs as a cause of low back pain with sciatica, interest in myelography was intensified, and numerous reports followed the pioneer work of Mixter and Barr. Lipiodol myelography lost some of its handicaps when Kubick and Hampton described their method for removal of lipiodol through the lumbar puncture needle used for its introduction.

The radiologic diagnosis of herniated discs soon was recognized to depend mainly on proper myelographic evidence. However, the adverse reactions to lipiodol remained uppermost in the minds of many observers, and the use of air myelography again was suggested. In a comparison of air and oil myelography, Barr and Mixter observed in 1941 that the diagnostic accuracy of the former was between 50 and 60 per cent compared to over 90 per cent for lipiodol. Together with removal of lipiodol by aspiration, the increased accuracy of this technic made it the one of choice.

In 1942 Strain, Plati and Warren reported a new contrast material for myelography. This preparation, ethyl iodophenylundecylate, was less viscous than lipiodol, flowed smoothly, provided excellent contrast and at first was believed to be absorbable. The first three qualifications were sustained by later work, but its absorbability proved to be slight or nonexistent. Nevertheless, ethyl iodophenylundecylate (pantopaque) is a superior contrast material, and its constant use since 1942 has established it as the best agent now available for myelography. Together with adequate removal after proper fluoroscopic and film examinations, it provides an accurate and well tolerated procedure. It should be realized that untoward reactions to pantopaque have been reported. Even though these are infrequent, and although many examination have been done with the pantopaque left in the spinal canal, the removal of the contrast agent should be a routine part of the procedure. As much as possible should be removed, recognizing that in some patients and under certain conditions removal of the entire quantity is not possible, and that small quantities remaining in the canal have not proven to be of any significance clinically. In a relatively small percentage of cases the examination may be followed by severe headache, much the same as occurs after diagnostic spinal puncture. A milder type of headache is somewhat more frequent. Occasionally the patient will complain of some increased lumbar pain or a stiff neck after myelography, but this usually subsides within a few hours. The question as to whether the inflammatory changes associated with myelography are due to the contrast agent or to the incident trauma or some break in proper technic is a fair one, and I am inclined to attribute such reactions more to the latter suppositions. This statement is based on the fact that about half the cases of arachnoiditis seen by us were in patients who had lumbar punctures without myelography, and in recent years we have seen three patients who had arachnoiditis following myelography after which almost all of the pantopaque had been skillfully removed.

Myelography is a radiologic examination, and should be done by well trained, careful radiologists who are aware of the indications for the investigation, who have evolved a reliable and critical set of criteria, and who can by virtue of their experience and ability accept the technical and interpretative responsibilities inherent to the procedure. Myelography should not be undertaken to satisfy diagnostic curiosity, but should be regarded as a necessary step to map out conditions which perhaps are correctable.

I have heard remarks, sometimes from highly regarded colleagues, to the effect that the examination is so fearsome that laminectomy is preferable. This is an unrealistic and unwarranted attitude. If properly performed myelography produces no more discomfort than a well done lumbar puncture. In fact, I do not hesitate in selected cases to perform the examination with no other preparation than a drop of novocaine subcutaneously. I have often mentioned to these patients that it would take about twenty minutes for the anesthesia to take effect, and while waiting I would check them fluoroscopically. During that time the examination was completed and the pantopaque removed without the patient being aware that the study had been performed.

The quantity of pantopaque necessary for myelography has received considerable discussion. It was recognized relatively early that about 5 to 6 c.cm. was a reasonable quantity for most purposes, and that larger amounts might be required for cervical examinations. Certain investigators recommended quantities up to 30 c. cm., mentioning that this made it easier to remove the pantopaque and facilitated the study. I believe that the quantity of pantopaque necessary depends much on the patient and his problem. If a complete block is suspected, as little as 0.5 c. cm. suffices. If the patient has a capacious canal considerably more may be required and I have used as much as 30 c. cm. in occasional cases. Usually the quantity should be that which fills the canal up to the level of the fourth lumbar interspace when the patient is erect.

Pantopaque usually is introduced into the third lumbar interspace by means of a 17 or 18 gauge needle, inserting the tip with the bevel directed parallel to the long axis of the spine so that the point passes through rather than cuts the longitudinal fibers. This is done with the patient sitting because I can get a midline puncture better than with the patient in the conventional horizontal flexed posture. Pantopaque is not instilled unless the flow is free. If there is any question as to the position of the needle point films are made in the posteroanterior and lateral decubitus views before proceding. When necessary 0.1 c. cm. of pantopaque is instilled and checked for configuration and movement in the canal. The examination is conducted on a table capable of tilting 90 degrees in either direction, with adequate support for the shoulders and feet. Emphasis is placed on following the pantopaque column fluoroscopically, and spot films are taken as necessary. The importance of following the head of the column rests in the fact that small defects and central veil-like lesions can best be detected by changes in the passage of the column over the lesion. The spot films are made in the posteroanterior and lateral views and in varying oblique projections as indicated by fluoroscopic study. Transabdominal views are important for investigating the anterior aspect of the column . If there is a question as to the configuration of the dorsal aspect of the pantopaque column, larger quantities may be required. The possibility of indentations into the spinal canal from the inferior aspect of the neural arches or the ligamenta flava may require relatively large quantities of pantopaque and filming of the patient in flexion and extension in the erect posture helps bring out these deformities. The examination should include bringing the column well up above the suspected area, and we routinely study the cervical area even if the suspected lesion is in the lumbar canal. This is especially important when lumbar myelography is normal and the patient has signs referable to the ventral and ventrolateral tracts.

Cervical myelography requires investigation of the spinal canal up to the

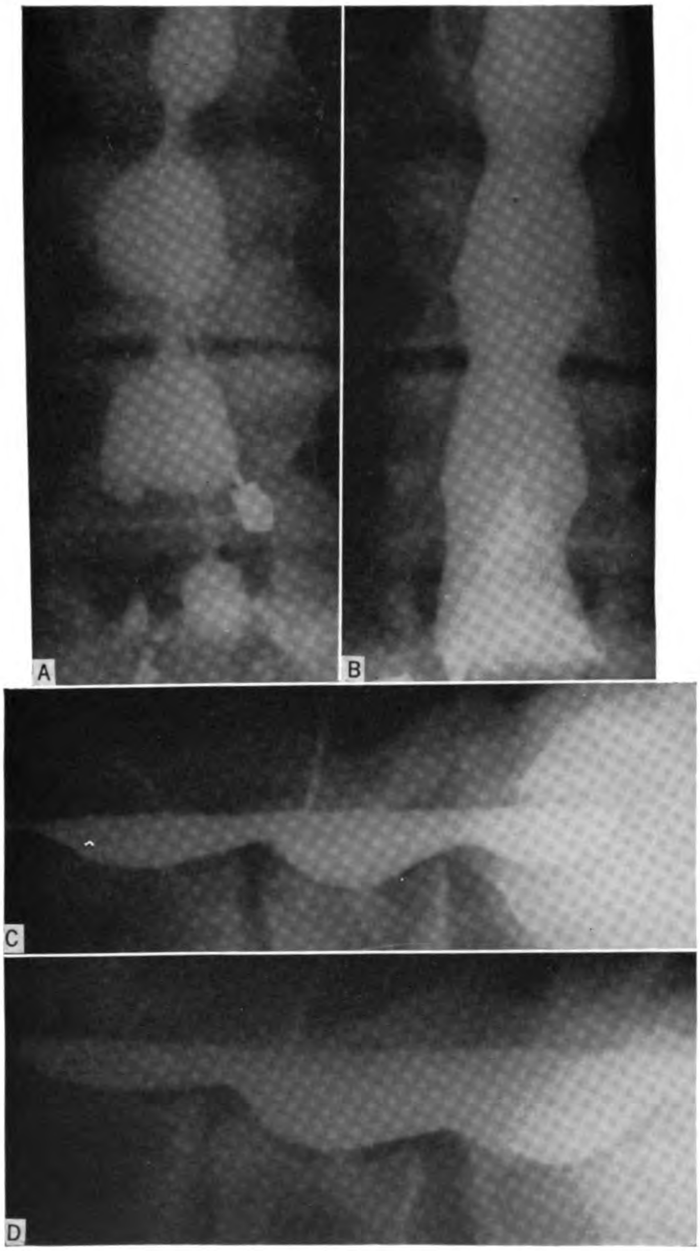


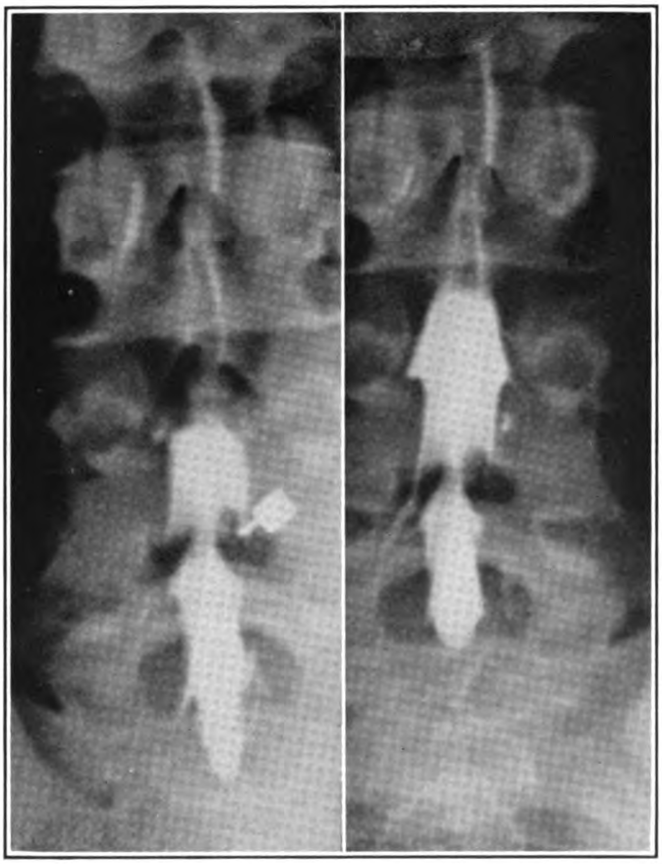
FIG. 42.—*A*, Anteroposterior myelogram, 6 cc. of pantopaque. Note the multiple hour-glass defects. The one at the narrowed third lumbar interspace is of particular interest.

B, Same patient, 18 cc. of pantopaque. Note that the defect at the third lumbar interspace is eliminated, as are those above and beneath.

C, Same patient, 6 cc. transabdominal examination. The pantopaque fills the concavities in the dorsal aspect of the vertebral bodies. Indentations at the level of the intervertebral discs are responsible for the hour-glass deformities of the column, attributed to the large capacity of the spinal canal.

D, Same patient, 18 cc. transabdominal examination. After adding an adequate quantity of pantopaque, the defects at the intervertebral discal levels are no longer present. This was a capacious canal and proper examination required larger amounts of pantopaque.

cisterna magna. With proper coordination it is possible to outline the cisterna magna and the cisterna pontis without introducing undue quantities of pantopaque into the lateral basilar subarachnoid spaces. This should be done whenever a lesion at the foramen magnum is suspected. The use of pantopaque myelography for investigation of the fourth ventricle and the aqueduct of Sylvius has been suggested recently (Mones and Werman, 1959). It is of historic interest to note that such possibilities had been contemplated by the early workers in the field, notably Sicard and Forestier and Peiper and Klose. Incidentally, the latter authors quoted Sicard and Forestier as remarking that lipiodol injected intracisternally could be removed by lumbar puncture.



A *B*

FIG. 43.—*A*, Anteroposterior myelogram showing defect at interspace between fourth and fifth lumbar vertebræ, with needle *in situ*. The question of needle defect had to be excluded. *B*, Same case with needle removed. Defect persisted. Operation disclosed herniated disc in midline.

Certain changes in the pantopaque column are misleading. Among these is the so-called "hour-glass" defect, usually observed in the lumbar and lower thoracic areas. This is due to the fact that pantopaque gathers in the hollows of the dorsal aspect of the respective vertebræ so that the upward bulge of the intervertebral discs indent the column and in some instances separate the pantopaque into pools. This is seen when relatively small quantities of pantopaque are used in wide spinal canals. This question can be resolved quickly by increasing the amount of pantopaque and positioning the column over the suspected area in adequate volume. Defects also have been ascribed to the placement of the lumbar puncture needle.

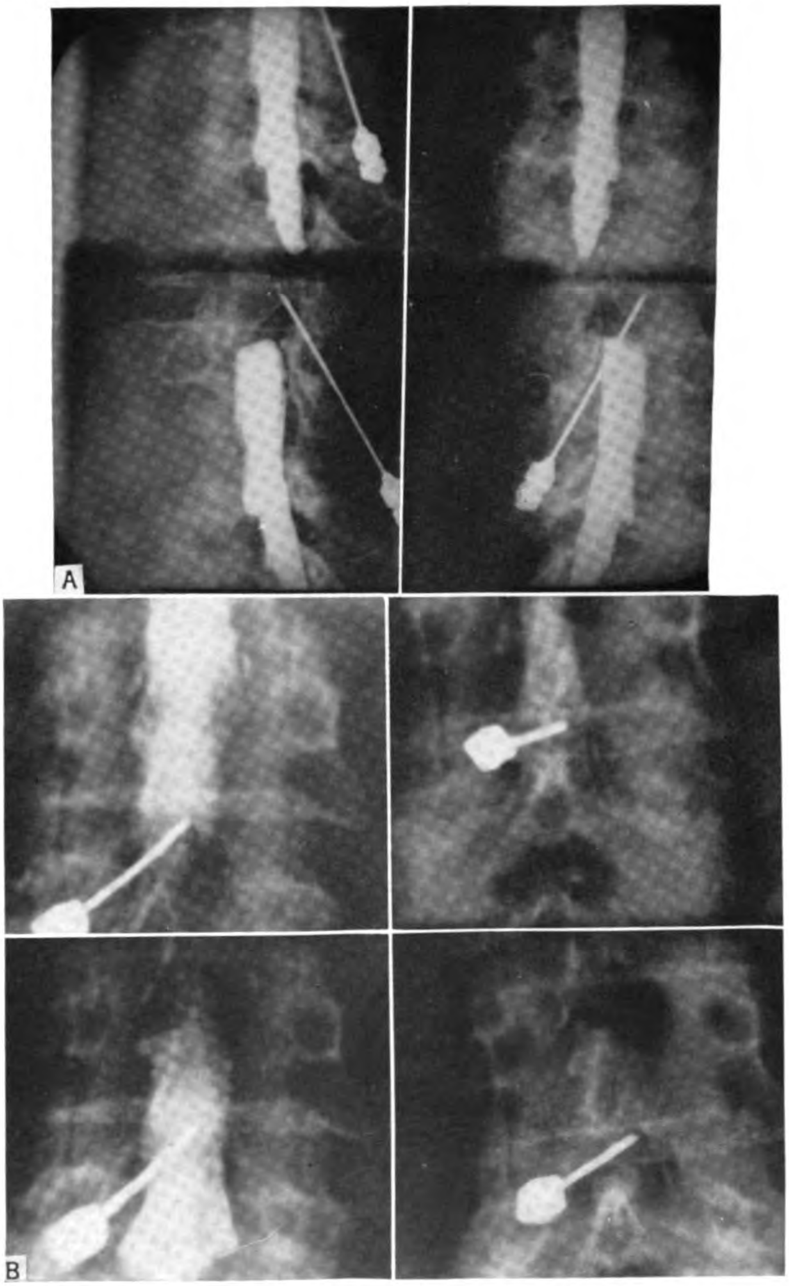


FIG. 44.—*A*, Spot roentgenograms in oblique projections which document fluoroscopic observation. *B*, Spot films made during removal of pantopaque. Note that even the fragmented pantopaque globules can be removed.

The explanation for the "needle defect" is that some cerebrospinal fluid enters the subdural space, resulting in a marginal indentation. I regard this as an overrated phenomenon. When one considers how many myelograms are done with the needle in situ and how few are the associated defects, the possibility assumes less awesome proportions. No defects appear when the needle tip is in the proper midline position. However, when the tip is placed laterally it may transfix the dural sheath and by causing some inward displacement could conceivably produce a defect. If this is suspected, the needle should be removed and the examination continued. My own experience has been that defects which persist have been associated with discal lesions. Another tap is required for removal of the pantopaque, preferably at a lower interspace.

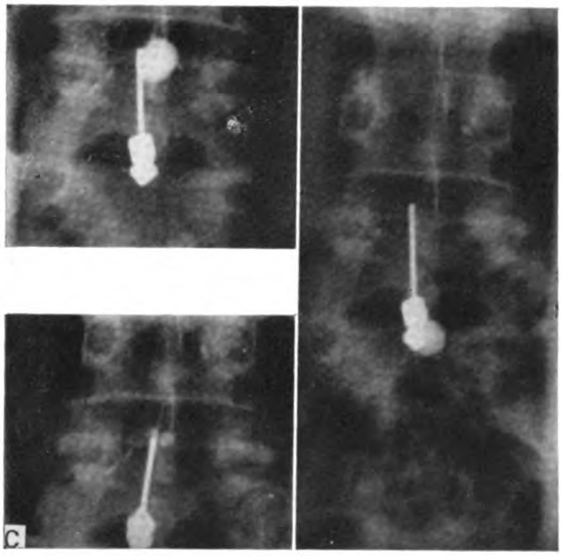


FIG. 44*C*.—Spot films made during removal of last globule of pantopaque. Note the thin filaments of oil persisting in the axillary pouch areas just beneath pedicles.

The configuration of the caudal sac should be carefully observed, particularly its width in relation to that of the spinal canal and its point of termination. When the sac terminates in a high position, or when it is relatively narrow in relation to the interpedicular space, laterally placed discal herniations may well exist without causing visible defects.

After the fluoroscopic part of the examination has been completed, the films reviewed and adequate cross-table exposures made, the pantopaque is removed. If the needle point is well centered, it is frequently possible to carefully and slowly aspirate most or all of the pantopaque after pooling it about the needle tip. Prior to removal the position of the needle tip should be ascertained from the cross table views. If the tip is only half way down the canal it is probably that only fluid or fluid mixed with pantopaque will be obtained. The removal of the pantopaque is greatly facilitated by placing the needle point so that it rests at the bottom of the spinal canal. Fairly often a nerve root or a bit of dura will occlude the needle opening, and aspiration then only serves to irritate. The needle point may be turned and aspiration again gently attempted. If difficulty is again encountered in getting a good flow, it has been found useful to get the patient to strain or cough. This often results in expulsion of the pantopaque without risking trauma to the nerve roots or dura, and good evacuation can be obtained with this maneuver alone. Occasionally it is good judgement to withdraw the needle and replace it lower down rather than traumatize the area by injudicious aspiration. When the pantopaque column breaks into droplets towards the end of removal, it becomes difficult to aspirate the last few drops even with repeated fluoroscopic localizations. One should not make the mistake of trying too hard to get these droplets because of the increase in fluoroscopic exposure and possible irritation of nerve roots.

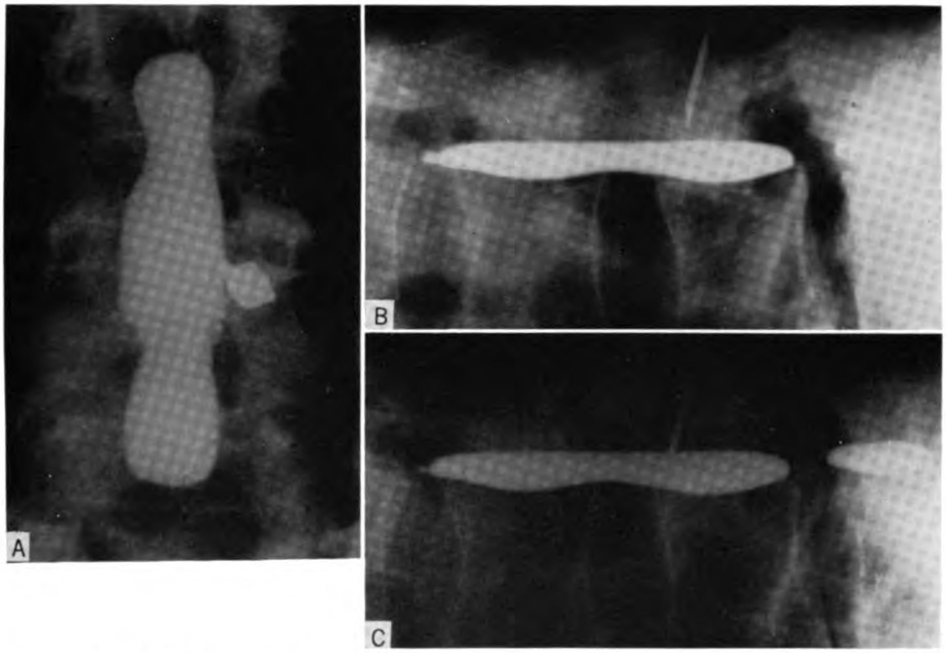


FIG. 45.—*A*, Myelogram with 6 cc. of pantopaque, patient in supine position. Cerebrospinal fluid aspirated. *B*, Same patient, transabdominal view. The needle tip is well above the pantopaque, so that mainly fluid was withdrawn. *C*, Same patient, with needle tip lowered into pantopaque pool, permitting its aspiration.

Pantopaque may escape from the subarachnoid region into the epidural space if the lumbar tap lacerates the dura, or if a puncture has been performed a day or two preceding myelography. In such an instance the contrast medium may pass high into the epidural area, outlining the space as high as the cervical canal and entering the nerve sheaths for variable distances. While this presents a rather dramatic picture, it is not associated with clinical symptoms. The pantopaque is slowly absorbed, and several patients followed over a two year period showed disappearance of most of the contrast medium with no untoward effects. If the needle tip is so situated that part of the bevel is outside the subarachnoid space and part the subdural space, pantopaque may enter the subdural region, producing a rather characteristic picture. The pantopaque passes slowly around the spinal canal as the patient is tilted, the flow being much less rapid than normal. As the patient is returned to the erect position the subdural pantopaque remains as a rather light tubular column while the subarachnoid pantopaque drops to the caudal sac. There is usually no clinical sequelæ to this occurrence.

The entrance of pantopaque into the venous circulation is a rather infrequent complication. Intravasation occurs during the injection, or if the patient should strain violently, or cough during the examination, in such

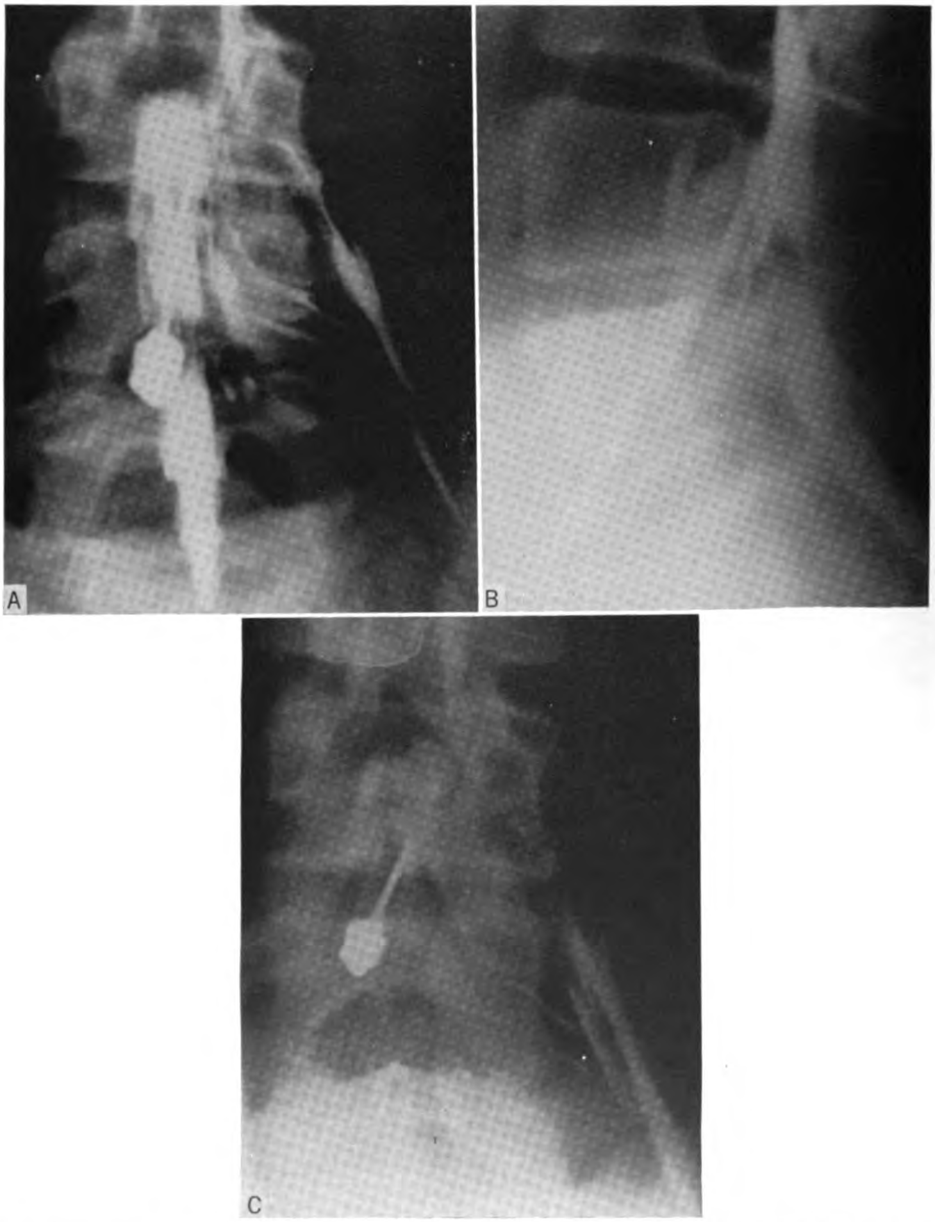


FIG. 46.—*A*, Extradural extravasation of pantopaque along third and fourth left lumbar roots and along left side of canal. *B*, Same patient, lateral view. The defect at the fourth lumbar interspace was caused by a large herniated disc, confirmed at operation. *C*, Same patient after removal of pantopaque. The intrathecal pantopaque has been removed almost completely, but that in the epidural spaces persists.

a manner that a vessel is torn by the needle tip. Serial films may demonstrate filling of the vertebral and paravertebral veins if one is quick in recording the occurrence. In the case I observed the oil suddenly assumed a bursting star-like configuration and vanished during the few moments of examination, passing from view along the paravertebral veins. Pulmonary embolism follows, but other than transient chest pain, mild hemoptysis and cough, and occasionally mild fever, no ill effects have been reported from this accident. Pantopaque injected inadvertently into a vertebral body causes a similar picture, easily understood as a form of intraosseous angiography.

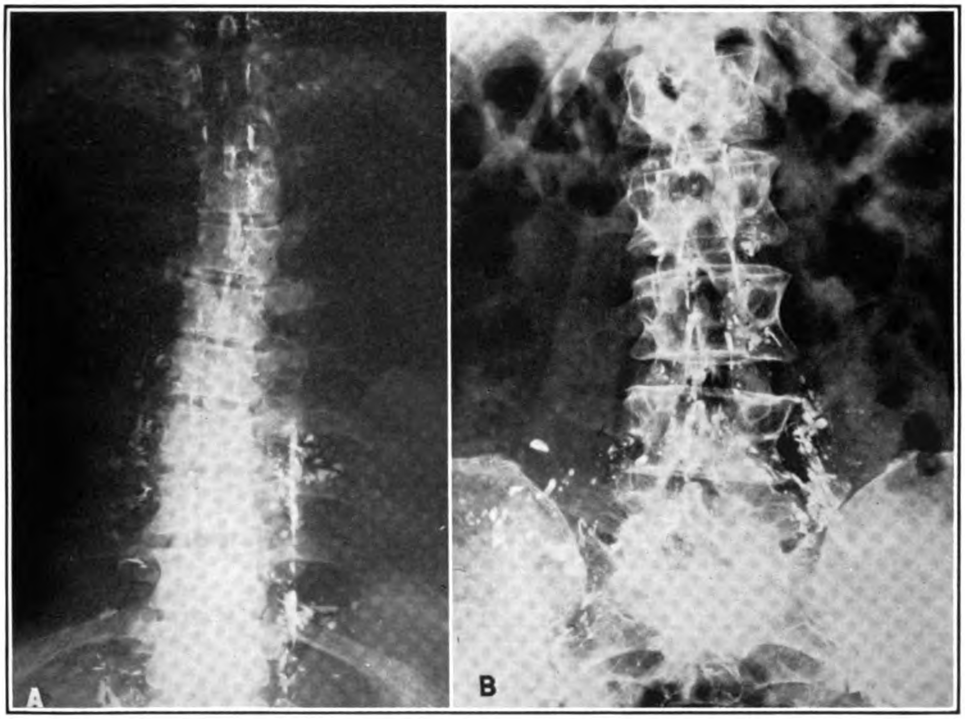


FIG. 47.—Epidural extension of pantopaque in cervical, thoracic and lumbosacral regions. Film taken three months after myelography.

Other effects of myelography have been associated with the inflammatory consequences of intrathecal injection of contrast materials. This has been well substantiated in the case of lipiodol. However, it appears that pantopaque has a lesser degree of inflammatory potential, and one cannot be accurate in deciding whether the inflammation results from faulty technic or the contrast material only. Arachnoiditis has been reported as a consequence of myelography, but it is only fair to reflect that many cases occurred in patients who had diagnostic lumbar punctures, intraspinal anesthesias, intrathecal introduction of medication or subarachnoid bleeding from various causes.

The effect of the entrance of pantopaque into the cranial cavity cannot be assessed accurately. Nevertheless, unless there is a valid necessity for outlining the basilar cisterns with pantopaque, I prefer to use air first. It is possible with proper timing of the positioning of the patient under fluoroscopic control to pass the pantopaque into the cisterna magna and to recover it almost completely. The control of the medium entering the cisterna pontis is not as complete, and the possibility of pantopaque entering the lateral cisterns from which it cannot be removed is greater when the cisterna pontis is filled. While the possibilities of inflammatory reactions in this area are slight, they have been reported. Schurr, McLaurin and Ingraham described an arachnoidal inflammatory reaction after injecting pantopaque into the subarachnoid spaces of dogs. Even though the quantities used were greater than employed clinically, the reaction indicated the desirability for preventing the entrance of pantopaque into the cranium. Erickson and van Baaren reported a 39-year-old man who expired fifteen months after myelography because of an exudative arachnoiditis obstructing the basilar cisterns and the fourth ventricle.

A more common concomitant of myelography is low back pain, occasionally with aggravated sciatica, headache and meningismus. These may be due to the lumbar puncture itself, or to vigorous attempts to aspirate the pantopaque. As a rule these symptoms disappear relatively quickly. Delayed reactions, in which severe headache and cervical stiffness develop two or three days after the examination, are occasionally encountered and sometimes can be quite distressing. These, too, disappear after periods of from a few hours to as much as five or six days and leave no sequelæ. Relief can be obtained by keeping the patient recumbent. Often the observation has been made that no discomfort is felt while the patient is lying down, but the headache becomes severe as soon as the erect posture is assumed. It is advisable to restrict activity until the patient is able to walk about with no discomfort, preferably waiting for about a day before resuming activities. In some patients operated upon within a day after myelography symptoms present just before surgery are absent soon thereafter.

Rare complications include extraocular muscle paralysis as reported by Wright. Pantopaque meningitis secondary to hypersensitivity has been described by Luce, Leith and Burrage.

Another question which arises frequently concerns the accuracy of myelography. Myelography, like other technical diagnostic procedures, is subject to individual variations both in elegance of performance and interpretation of observations. It is difficult to do more than get a broad idea of what can be accomplished. In my own experience the accuracy has been about 90 per cent. Published figures vary from about 65 per cent to a high of 96 per cent. The accuracy appears greater when the examinations are conducted by one individual, when the patients are carefully selected, and when the clinical problem is clearly understood before proceding.

As one reviews the literature it becomes apparent that not all are in accord with the opinion that myelography should precede exploration of the spinal canal. In 1941 Dandy, in a paper entitled "Concealed Ruptured Intervertebral Discs" included in the heading "A Plea for Elimination of Contrast Examinations in Diagnosis." Derry (1943) believed that physical examination was sufficient to indicate the level of a lesion, and that contrast examinations were unnecessary, an opinion echoed by O'Connell (1951) and others. Myelography was advocated by some in selected cases when the clinical picture was confusing (Key, 1952). The dangers of overlooking spinal lesions because of reluctance to use myelography was emphasized by Svien, Dodge and Camp (1951). These authors reported an analysis of 1245 surgical cases of herniated lumbar disks and 51 cases of spinal cord tumors which arose at or below the tenth dorsal vertebra. They found that in twenty cases of spinal cord tumor the preoperative diagnosis had been herniated disk. In seven others the possibility of a herniated disc or spinal cord tumor had been considered. The signs and symptoms in all 27 cases strongly suggested discal lesions of the lower two lumbar interspaces, but in only 5 were the tumors located there. In the other 22 the lesions were cephalad to the fourth lumbar level.

I am convinced of the importance of myelography before exploration of the spinal canal, and regard it in the same light as a gastrointestinal radiologic examination before operation for a stomach lesion. While not infallible, it compares very favorably with other diagnostic procedures, is well tolerated, has relatively little ill effect, and can contribute much to accurate diagnosis.

THE NORMAL MYELOGRAM

Gas myelograms delineate the lumbar spinal canal as a smooth-walled radiolucent tube. The cauda equina nerve roots cannot be identified. In the thoracic canal the cord, enveloped by gas, is identified as a shadow of relatively increased density. Its delineation, especially in the lateral projection, is enhanced by body section radiography. Cervical gas myel-

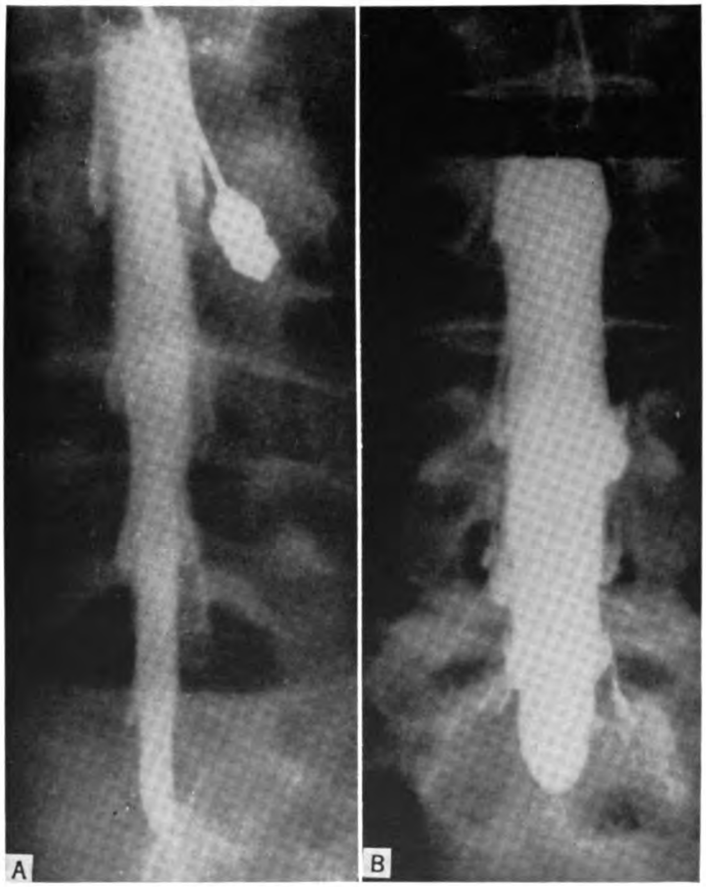


FIG. 48.—*A*, Narrow caudal sac. Six cc. of pantopaque have filled the sac up to the level of the third lumbar vertebra. Erect examination. *B*, A wide spinal canal, erect examination using 24 cc. of pantopaque.

ography likewise identifies the spinal cord by replacement of the surrounding cerebrospinal fluid, and is well suited for demonstration of the cerebrospinal fluid spaces in the cervico-occipital area. It is worth checking this region routinely on lumbar pneumoencephalograms, particularly if there is any possibility of a lesion which may project downward from the foramen magnum.

Because gas can be introduced readily, and is completely absorbed in a relatively short time, the examination is done with facility. It is more suited for the identification of gross lesions such as tumors or advanced spinal cord atrophy. Its reliability in the diagnosis of discal herniations is not sufficient to warrant its use, and its inability to visualize the axillary pouches in the lumbar spinal canal is a handicap. Complete replacement of cerebrospinal fluid with gas affords the maximum information available from this method, and in addition may be utilized for pneumoencephalographic investigation if desired.

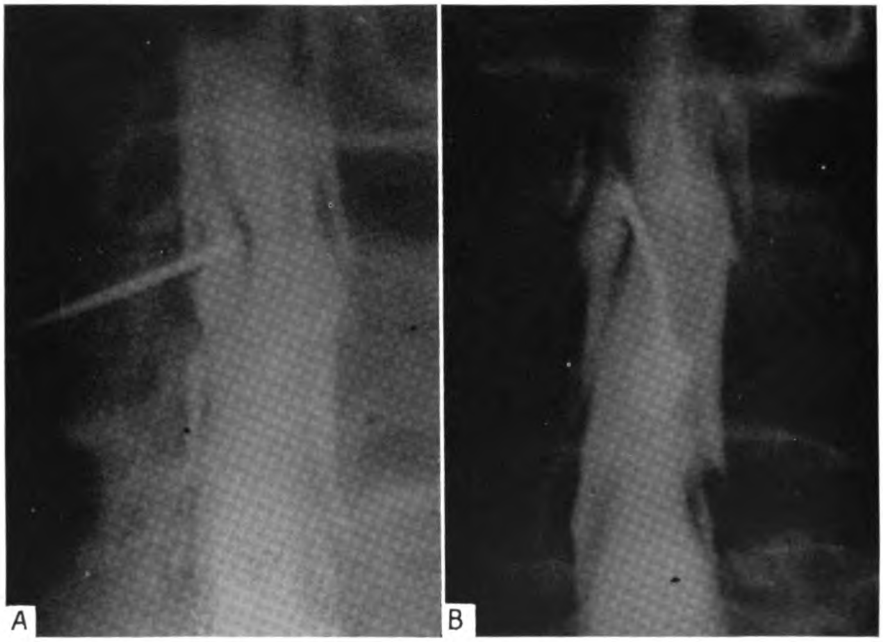


FIG. 49.—Deflection of cauda equina roots by needle tip. There was no complaint of pain when this was present.

The normal lumbar caudal sac tapers to a point usually at the level of the inferior aspect of the fifth lumbar interspace or somewhat lower. Occasionally a thin streak of pantopaque is seen extending further caudad along the termination of the filum terminale. The relative width of the caudal sac should be checked in relation to that of the fifth interpedicular space. If the sac is less than 16 mm. in diameter, or if the interpedicular space is wide, lateral discal lesions cannot be anticipated to produce visible impingement on the pantopaque column, and may not even affect the axillary pouch. Quite often the caudal sac terminates as a relatively wide, broad structure terminating at the first or second sacral levels. As a rule, this is not significant. However, these variations should be differentiated from occult meningoceles, which will be discussed later.

The axillary pouches can be identified on either side of the dural sac. These nerve sleeves are of variable length, and usually do not excede 1 to 1.5 cm. in length. The fifth lumbar root passes through the intervertebral foramen between the fifth lumbar and first sacral vertebræ. Occasionally the first and second sacral roots are visible beneath the axillary pouch of the fifth lumbar nerve. The axillary pouches represent the tubular envelopment of the dural sheath as it merges with the perineurium, and are directed obliquely downward and laterally. As a rule the lowermost three lumbar axillary pouches are more prominent than those above. Each pouch represents the nerve root corresponding numerically to the cephalad vertebra.

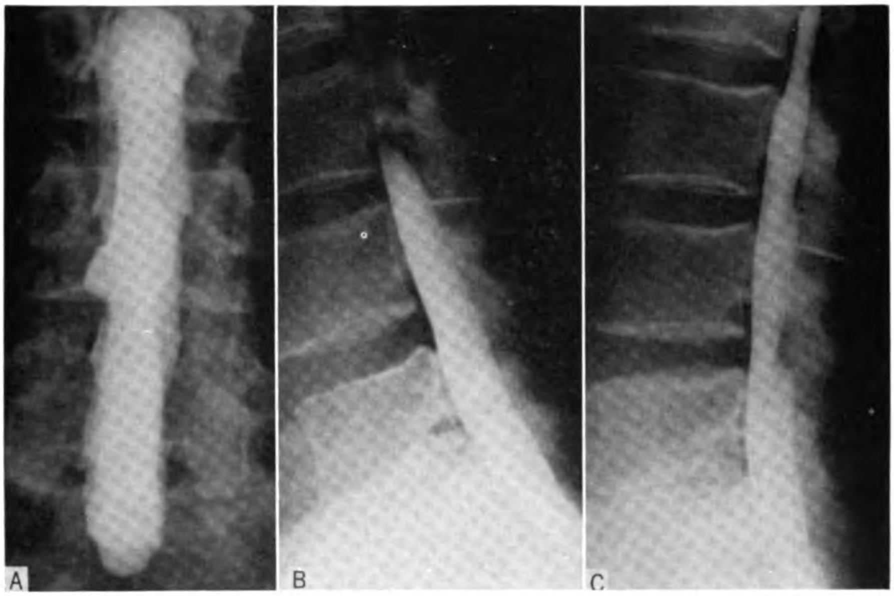


FIG. 50.—*A*, Erect large volume myelogram filling the lumbar canal to the level of the upper aspect of the third lumbar vertebra. *B*, Same patient in flexion, lateral projection. Note the smooth aspect of the dorsal as well as of the ventral aspects of the canal. *C*, Same patient in extension. Indentations appear in the dorsal aspect of the column from bulging of the ligamenta flava. Under strain the pantopaque column moves upward.

In many examinations the cauda equina is visible to varying degree. Occasionally displacement of a root by the lumbar puncture needle can be demonstrated, and it is of interest that even when the needle is seen displacing the root the patient does not complain of pain. Edema of the nerve trunks of the cauda equina also renders them more apparent on myelograms.

The anterior aspect of the lumbar spinal canal is best seen on cross-table exposures. These cannot be duplicated by turning the patient on his side to a lateral horizontal position unless large quantities or pantopaque are used because lesser amounts descend to the dependent portion of the canal. Films made with the patient in a lateral position and the x-ray beam directed horizontally sometimes are used to delineate the lateral aspects of the canal, and aides in the identification of the axillary pouches. If information is desired about the dorsal aspect of the canal it is necessary to instill sufficient pantopaque to fill the subarachnoid space up to the desired level. Defects are brought out best by making erect lateral exposures with the patient in flexion and extension.

Usually the pantopaque column is in close proximity to the dorsal aspect of the vertebral bodies. Frequently gentle, smooth indentation is produced by normal bulging of the intervertebral disc. At the lumbosacral interspace there may be a varying degree of elevation of the distal caudal sac from the floor of the canal. Usually this is symmetric and smooth, and in itself is not of pathologic importance.

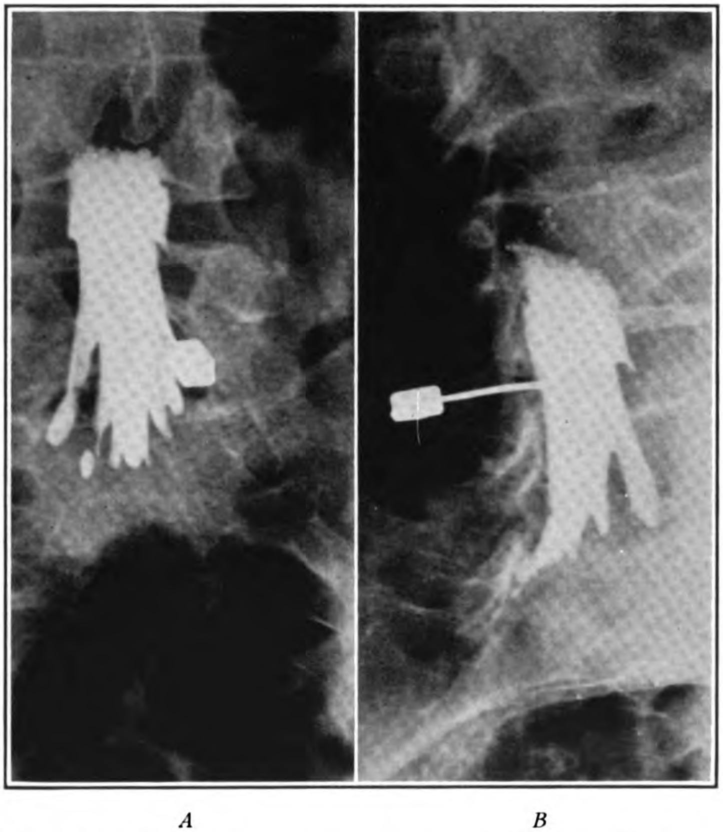


FIG. 51.—Myelogram with 6 cc. of pantopaque in anteroposterior and oblique views. The axillary pouches at the fifth lumbar and first sacral levels present arachnoidal diverticulæ. Note how the oblique projection brings out the deformity on the left side. No symptoms were attributed to these malformations.

Occasionally small diverticulum-like structures are seen filled with pantopaque, lying lateral to the axillary pouches. These are referred to as arachnoidal diverticula, or less correctly, as perineurial cysts. Sometimes they become large enough to cause nerve root compression or erosion of the adjacent bone, and under such circumstances assume surgical significance. They should be differentiated from the perineurial or extradural sacral cysts described by Tarlov as occurring at the junction of the posterior nerve root with the dorsal root ganglion, in the space between the perineurium and endoneurium as nearly solid masses containing fibers and ganglion cells. Since they do not communicate with the cerebrospinal fluid spaces they cannot fill with contrast material, and as a rule do not produce bone erosion unless quite large. Nevertheless, they can cause root compression symptoms. Arachnoidal diverticulæ sometimes are better demonstrated on films m de after a few hours if some pantopaque remains in the spinal canal because of delayed entrance of the contrast agent through a narrow sac. Pantopaque also may be retained in these structures for a considerable time, with no ill effects.

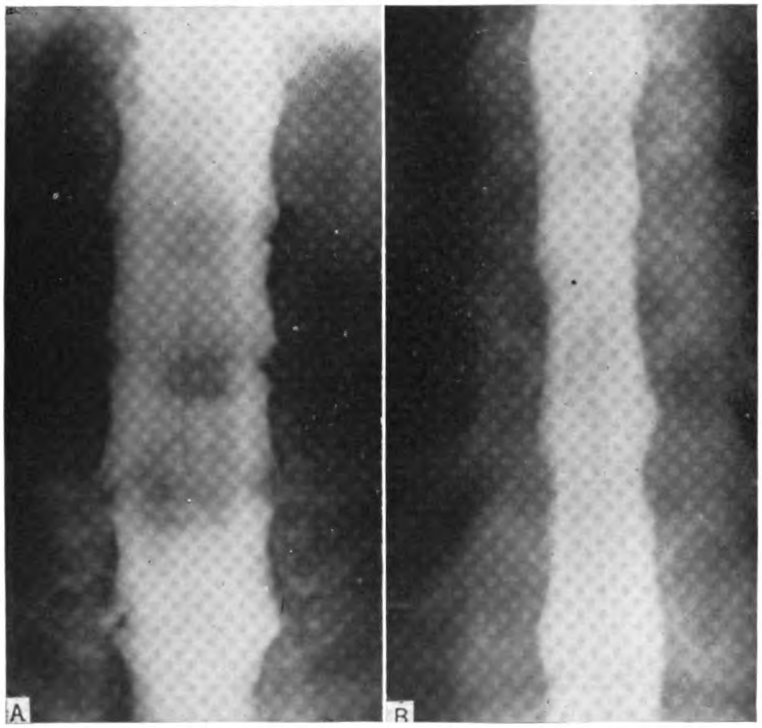


FIG. 52.—*A*, The anterior spinal artery is seen in the midline. The central radiolucency is caused by the spinal cord. Note the radicular arteries in the lower cervical region. *B*, Same patient. The anterior spinal artery is visible through the pantopaque column in the thoracic spinal canal.

The thoracic spinal canal as outlined with pantopaque presents a more slender appearance. The tubular configuration of the column often is difficult to maintain, and sometimes it is helpful to study this with the patient in the oblique or prone lateral positions. The nerve root sleeves rarely fill. Occasionally a thin linear radiolucent streak is visible in the middle of the pantopaque column, representing the anterior spinal artery. The density of the column reflects the relatively small space around the cord.

The cervical spinal canal is best studied when well filled. As a rule the pantopaque column meets a momentary delay as it passes the cervicothoracic junction, and the opaque medium first flows to either side leaving a radiolucent middle streak about 1.5 to 2.0 cm. in diameter, which represents the cervical cord. This should not be misinterpreted as indicative of a pathologic condition. The axillary pouches are directed almost hori-

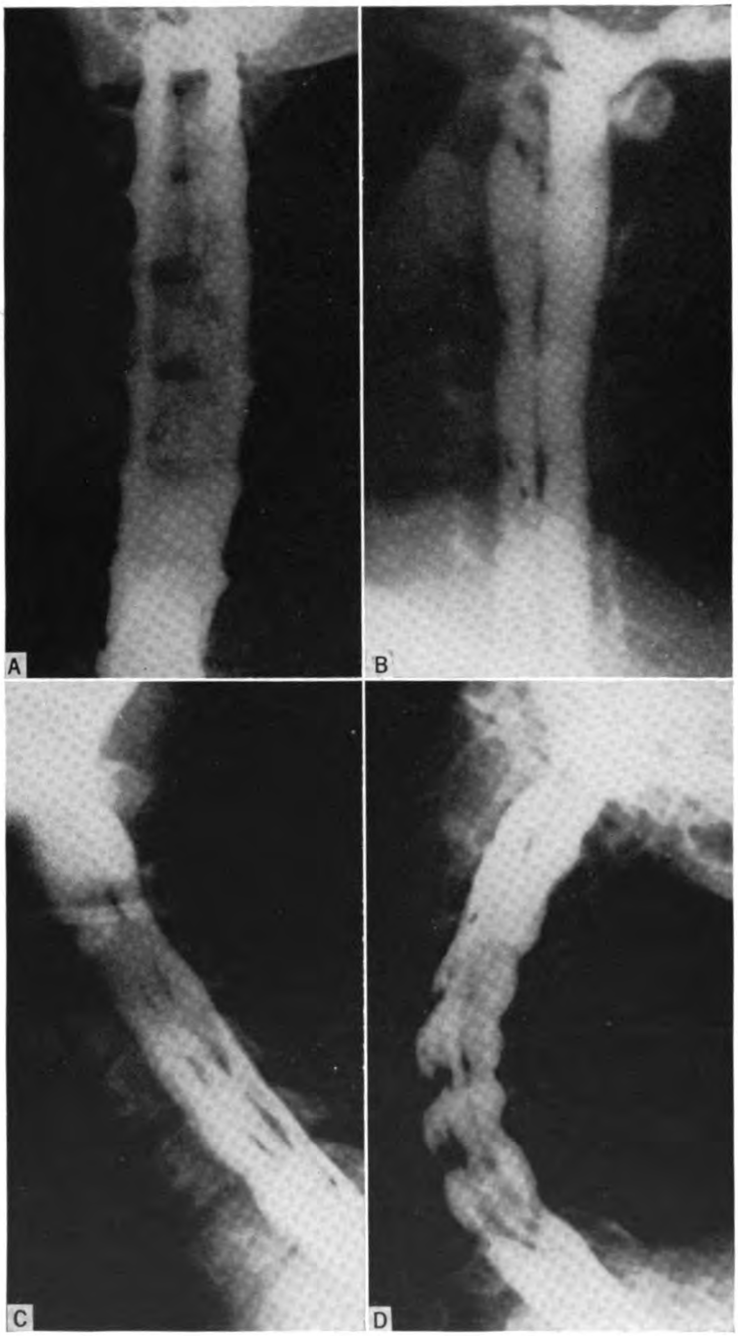


FIG. 53.—Cervical myelogram. A linear radiolucency is seen in the midline, produced by the anterior spinal artery. Several branches extend along the lower cervical nerve roots.

B, Transcervical myelogram, same patient. Pantopaque fills the cisterna magna. Note the radiolucent line in the middle of the column. This has been attributed to the dentate ligament.

C, Transcervical myelogram with head in flexion. Two radiolucent lines are present in the pantopaque column, possibly the dentate ligaments on either side of the cord. Note the smooth configuration of the dorsal aspect of the pantopaque column.

D, Same patient, head in extension. Note the indentations produced by the wrinkling of the ligamenta flava on the dorsal aspect of the pantopaque column.

zontally and fill quite readily. Turning the head from one side to the other permits one to fill the lateral spaces quite readily, but this is not necessary if adequate quantities of pantopaque are used. The axillary pouches sometimes show a radiolucent center for a short distance, produced by pantopaque on either side of the nerve root. It is important to obtain transcervical films with the patient prone, using a horizontal beam. Indentations along the anterior aspect of the column are best identified on this projection. If the canal is adequately filled indentations along the dorsal aspect may be

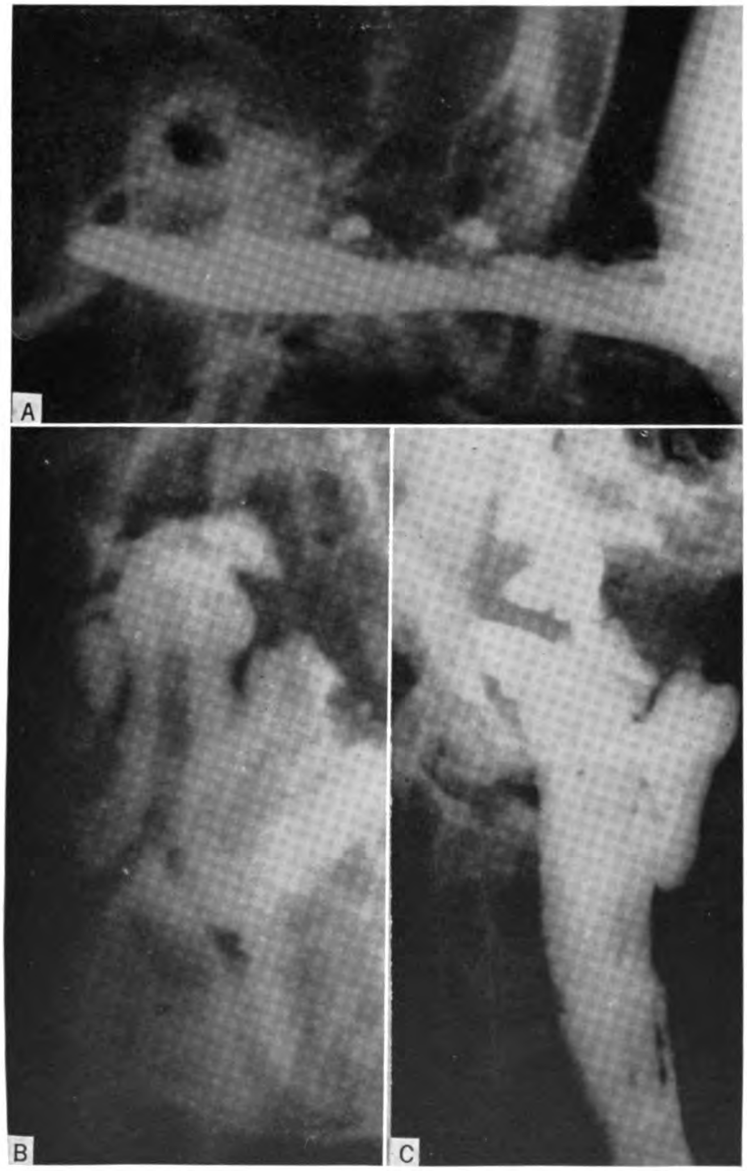


Fig. 54.—*A*, Transcervical myelogram with pantopaque filling the cisterna pontis. *B*, Same patient. The right and left vertebral arteries uniting to form the basilar artery present as radiolucent shadows displacing the pantopaque. *C*, Same patient, oblique view. The vertebral artery is seen at the level of the cervico-occipital junction, traversing the filled cisterna magna.

demonstrated. Infolding of the ligamentum flavum, which may have important clinical implication, are best identified by this maneuver, and exaggeration of the indentations are brought out by studies made with the head extended.

The cervico-occipital junction can be investigated without difficulty if the pantopaque is brought to this area under fluoroscopic control. Much depends on the skill and coordination of the examiner and the cooperation of the patient. The entrance of contrast material into the basilar cisterns laterally should be avoided. It is possible to introduce pantopaque into the cisterna magna and pontis, and to recover almost all of it if one acts quickly and does not permit the oil to spill laterally. If the pantopaque envelopes this area the vertebral and basilar arteries can be identified as radiolucent tubular shadows corresponding to these vessels.

When the cervical canal is well filled it is possible to identify the dentate ligaments, the thin fibrous bands on either side of the cord which separate the anterior from the posterior nerve roots. These appear on lateral studies as linear translucent shadows, the anterior and posterior surfaces being outlined by the contrast agent. The cervical dentate ligament is rather heavy, but the thoracic and lumbar dentate ligaments are less prominent and hence cannot be demonstrated. Teng points out that the continuity of these structures may be disrupted in cervical spondylosis. The anterior spinal artery sometimes is visible on frontal projections as a thin radiolucent streak in the middle of the canal, and occasionally branches passing to the axillary pouches are seen in the middle and inferior cervical region. When the upper cervical canal and the cisterna magna are well filled it is possible to outline the inverted Y-shaped junction of the vertebral and the basilar arteries.

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DISCOGRAPHY

Following experimental work in which he injected red lead anteriorly into the intervertebral disc, Lindblom applied his observations clinically for the demonstration of the normal and pathologic anatomy of these structures. In his experience myelography had proven somewhat lacking in accuracy, a deficiency he regarded as diminished by discographic investigations in which the three lowermost lumbar intervertebral discs were injected with water-soluble absorbable contrast material together with a small quantity of novocaine. At that time a mixture of 2 c. cm. of 35 per cent diodrast and 0.5 c. cm. of novocaine were used. The same quantities are still used, but at present hypaque is the preferred medium. Lindblom found that normal discs accepted about 0.3 c. cm. of the contrast medium, while pathologic discs take more.

The question as to the possible effects of puncturing multiple discs still causes concern. From the reports of Cloward, Fernström and others it would appear that the procedure rarely is complicated by infection. That infections can occur has been reported by De Seze and Levernieux, Scherbel and Gardner and Ectors. Earlier observations had indicated that trauma to the intervertebral disc might be accompanied by progressive narrowing, superimposed infection and consequent varying degrees of disability. This was accomplished experimentally in animals by Key and Ford and by Congdon. It has been maintained that the small bore of the needle used to penetrate the disc mitigated against the possibility of consequent damage, an opinion supported by the many reports of the successful use of discography.

The normal discogram reveals two contrast-filled spaces, one along the upper and the other along the lower vertebral margin, outlining the nucleus pulposus but not projecting into the annulus fibrosus. The two layers are joined by an irregular streak of contrast material through the center of the disc, presenting a "collar-button" appearance. Pathologic discograms show varying degrees of emergence of the contrast material, which may bulge into the spinal canal or demonstrate fissure-like passages laterally or anteriorly. The diagnostic value of discography is heightened because the patient's symptoms may be duplicated or heightened during the time the discal protrusion is distended. Contrast material sometimes passes through the ruptured annulus fibrosus and diffuses into the spinal canal. Lindblom (1951) mentioned a case in which the injection tracked anteriorly, causing severe abdominal pain. He also quoted Grill who directed attention to the possibility of abdominal pain arising from discal ruptures.

Discography has been credited with identifying a considerably higher incidence of multiple discal ruptures. In evaluating this, it should be recalled that many discal protrusions are asymptomatic and are best left alone. Gardner and his coworkers observed that when an intervertebral disc is narrowed the discogram is never normal, and that the nucleus pulpsus is disorganized. A high degree of intelligent clinical judgement is required to identify which protrusion is responsible for symptoms and which is not related to the patient's complaints.

Discography of the cervical spine has been advocated by Cloward, utilizing an anterolateral approach for the placement of the needles. Since most cervical disc pathologic change takes place in the lower two interspaces, Cloward advocates investigating these two areas. He regards this as a technic superior to myelography both in the lumbar and the cervical areas,

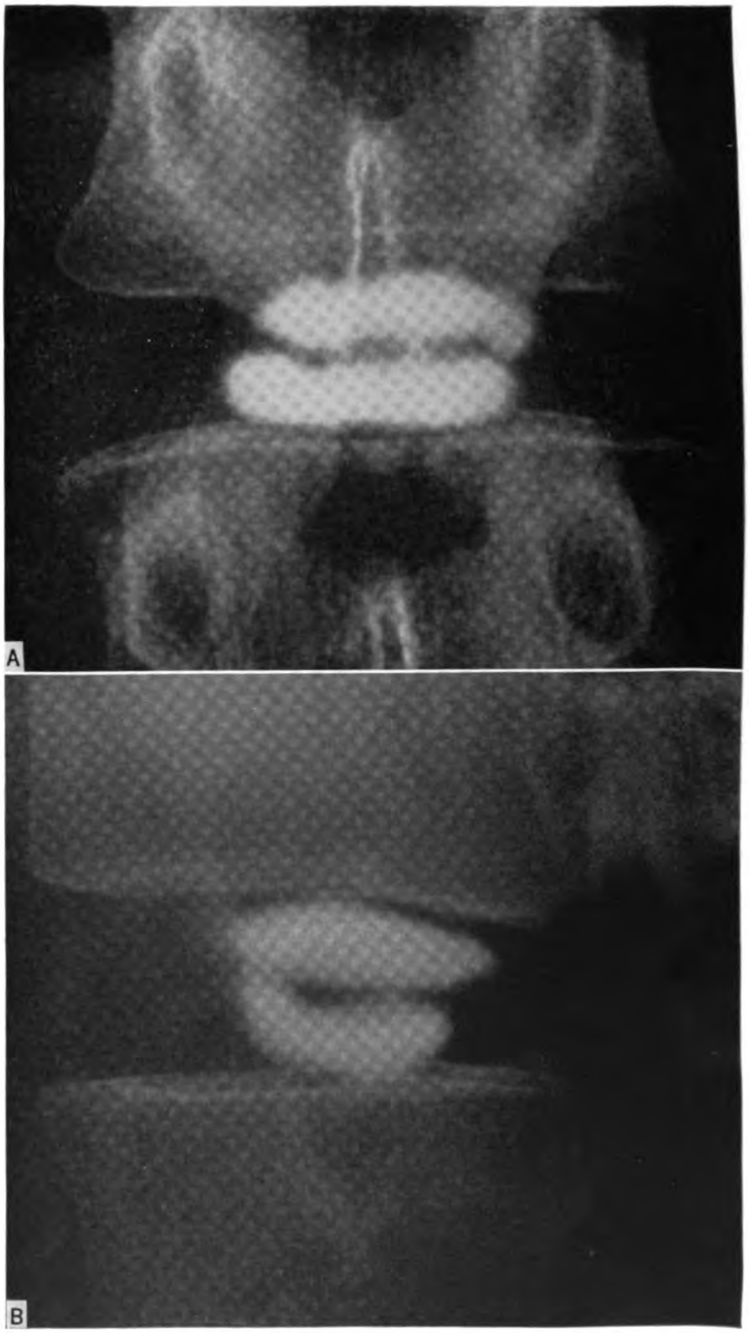

FIG. 55.—*A*, Anteroposterior and lateral (*B*) normal discograms. The contrast material collects in a double disc pattern. (Dr. Ulf Fernström, courtesy of Acta chirurg. scandinav.)

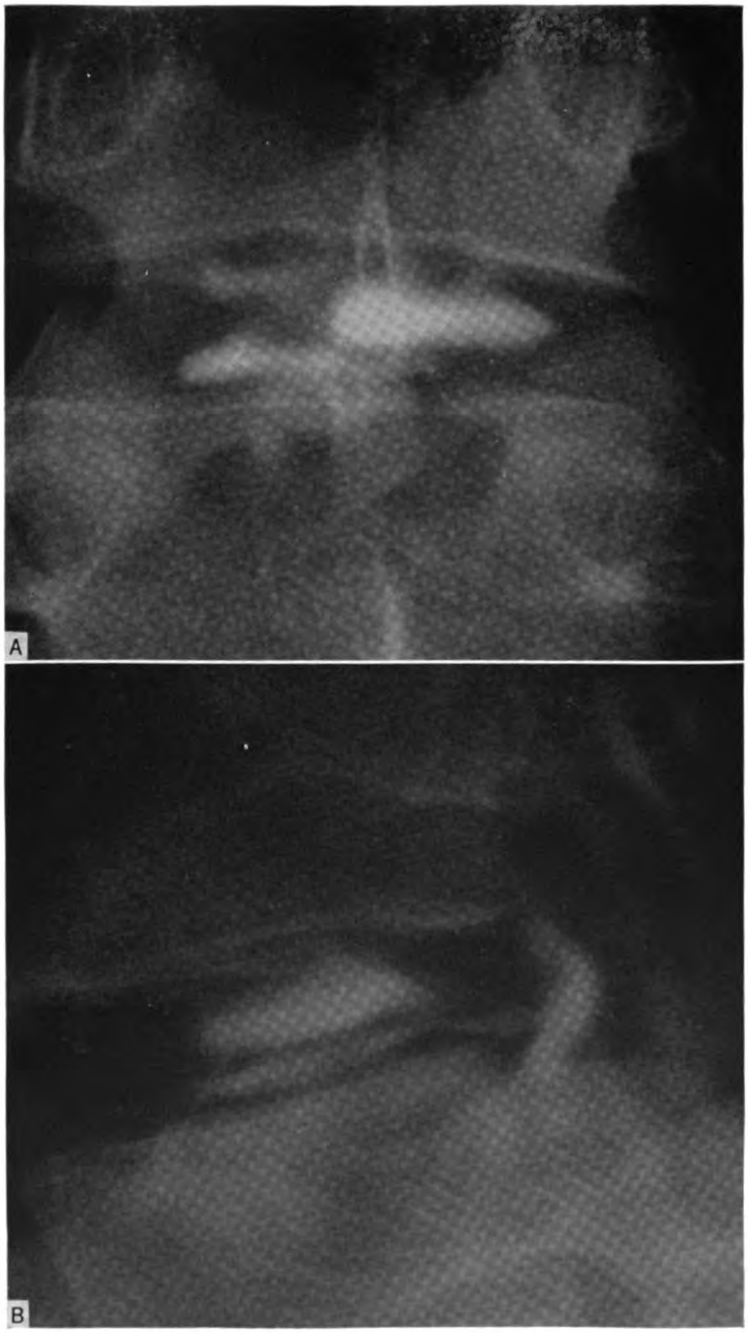

FIG. 56.—*A* and *B*, Anteroposterior and lateral discograms in a patient with a herniated lumbar disc. The contrast material extends to the area of herniation. (Dr. Ulf Fernström, courtesy of Acta chirurg. scandinav.)

providing in his opinion a much higher accuracy in diagnosis and a safer method of examination.

Even though there has been enthusiatic endorsement of discography as a safer, easier and more reliable method of investigation, one nevertheless must pause to consider and compare this procedure with myelography. There are many, including the present author, who hold that trauma of multiple discal punctures is not without hazard, and who feel that unless a definite superiority can be proved one should not abandon myelography. One of the disadvantages of discography is that it requires much manipulation for investigating limited areas. It seems almost self-evident that radiologic exploration of the spinal canal should not be limited to two or three discal interspaces. Also, the disclosure of an abnormal discal pattern in an adult patient may or may not have diagnostic significance pointing towards the need for surgical relief. This observation, by the way, is applicable to myelography. Nevertheless, the demonstration of definite multiple deformities myelographically has held up quite well on correlation of clinical and surgical observations. In our experience the number of patients with single lesions who have been relieved of their complaints after operation is vastly greater than those who have had multiple defects. Since a large number of these patients were in the middle age group, and who theoretically had considerable degenerative changes in the discs, the relief obtained by the correction of a single discal herniation is worth remembering when reviewing statistics of the multiple herniations identified discographically. Myelography also has the great advantage of permitting inspection of the entire spinal canal, and is therefore better applicable to the investigation for tumors. The inherent risks of the infective consequences of lumbar puncture would appear to be about the same whether myelography or discography is done, with the provisos that it seems reasonable that three punctures might be more hazardous than one. On the other hand, the introduction of contrast material into the spinal canal carries with it some risk as well.

In our experience myelography has proved sufficiently reliable for us not to employ discography for the diagnosis of herniated discs.

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Chapter

2

Malformations of the Spinal Column

DEVELOPMENTAL ANOMALIES OF THE OCCIPITAL BONE, ATLAS AND AXIS

The last occipital sclerotome sometimes forms as a partially separate entity, and becomes fused with the anterior margin of the foramen magnum, or appears as a bony nodule between the occipital bone, the atlas and the dens. This proatlas, or occipital vertebra, also may take the form of a partially separate entity, consisting of more than one bit of bone, and may distort the foramen magnum by rendering it irregularly narrow because of the appearance of accessory eminences along its periphery. Anteriorly, an articular condyle for the dens may be seen, and a partially formed hypocondylar arch completely or partly united with the anterior rim of the foramen magnum and a modified neural arch sometimes is present . Rudimentary transverse processes without foramina transversarii may be partly or completely united with the base of the skull.

Atlanto-axial fusion, or assimilation of the atlas and axis, results from maldevelopment of the caudal half of the last occipital and the cranial half of the first cervical sclerotomes. Complete fusion results in union of the first cervical vertebra with the foramen magnum circumferentially, while with partial fusion varying segments of the first cervical vertebra become attached to the foraminal rim. Most often the anterior arch becomes fixed. In the event of complete fusion, no symptoms appear. However, partial fusion may be associated with instability of the atlas and axis, abnormal mobility of the odontoid being produced by lack of fixation of the ligaments, particularly the transverse ligament. This abnormal range of motion can can be demonstrated on lateral roentgenograms in flexion and extension, and are better seen on laminagrams. Because of impingement of the bony structures on adjacent nerves and the spinal cord, a wide range of neurologic symptoms may appear, together with pain, abnormal attitudes of the head and limited ranges of motion. Neurologically these symptoms often mimic those of various degenerative diseases such as syringomyelia and multiple sclerosis. Weakness of the lower extremities and ataxia may be presenting symptoms, alone or together, with concomitant sensory disturbances. Often motor impairment of the upper extremities also appears.

A common deformity of the first cervical vertebra is failure of fusion of its neural arch, a relatively frequent and usually symptomless condition.

Differentiation of an occipital vertebra from atlantoaxial assimilation sometimes is difficult. With atlantoaxial fusion a foramen usually persists on each side between the posterior arch of the fused atlas with the adjacent occipital bone to permit passage of the vertebral artery and first cervical nerve, which retain their normal relations with the atlas and the base of the occipital bone. Atlanto-occipital fusion may be accompanied by fusion of two or more cervical segments. Accessory eminences occur on one or both sides of the foramen magnum, narrowing and distorting this area, much the same as occipital vertebræ. It has been stated that the two can be separated anatomically by observing the disposition of the articular facets. If the joint surfaces of a proatlas are projected as planes they will converge caudally as do those of normal occipital condyles. On the other hand, the articular surfaces of an assimilated axis will, if prolonged, extend cranially.

Ossiculum Terminale.—This anomalous structure occurs as a separate

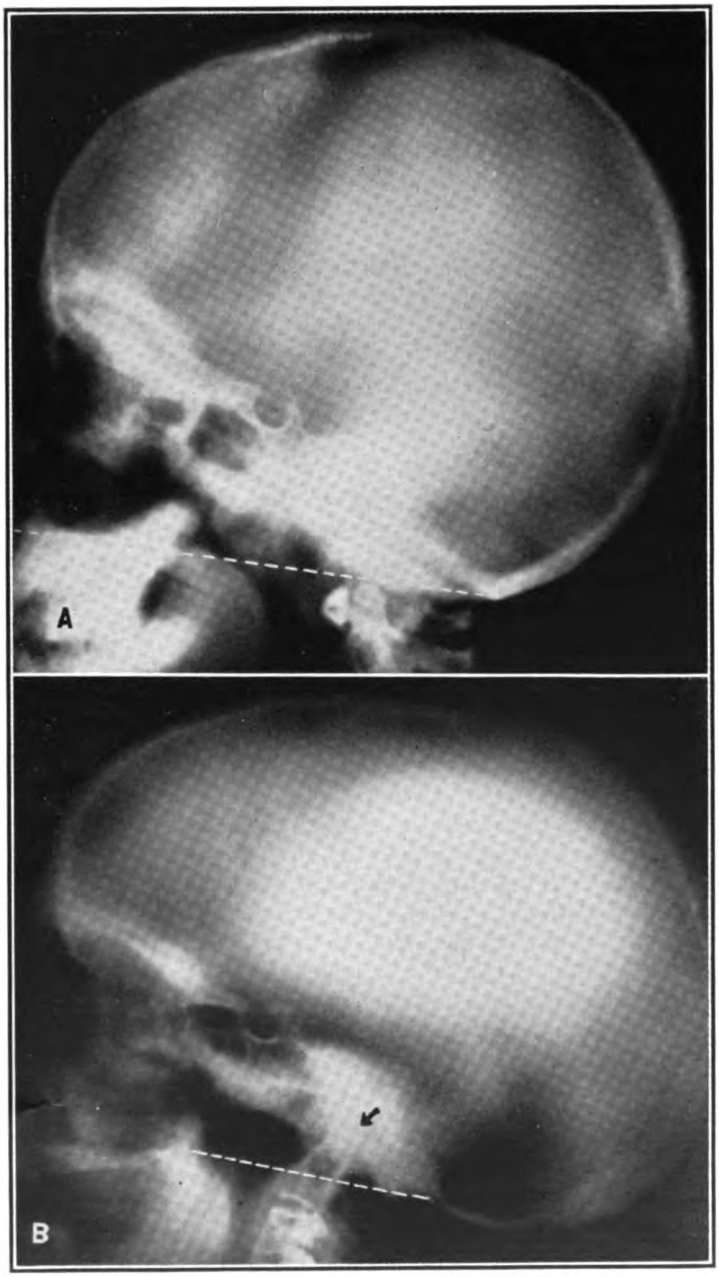

Fig. 57. Legend on opposite page.

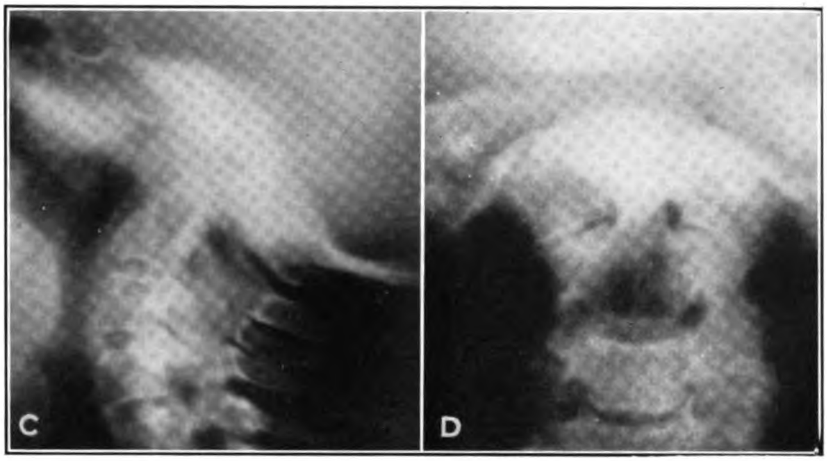

FIG. 57.—*A*, Lateral midline laminagram of normal skull. Chamberlain's line is indicated by the dotted white line. *B*, lateral midline laminagram, basilar invagination. Chamberlain's line passes through the base of the dens. The tip of the dens is displaced upwards (arrow). *C*, Same case, detail skull base showing upward convexity of base. The entire first cervical vertebra is displaced upwards and merges with the basiocciput. *D*, Same case, frontal body section roentgenogram. The occipital condyles and the lateral masses of the first cerviacl vertebræ are fused. Note the inward projection of the basiocciput.

ossicle which originates from a notochordal remnant in the terminal ligament of the odontoid process. In the normal subject this usually unites with the odontoid at about the age of six years. However, when its fusion is incomplete it may persist as a separate bone associated with a shortening and blunting of the dens. It should be distinguished from the so-called "third condyle" which develops from terminal or cervical ligaments as a tubercle on the anterior inner rim of the foramen magnum, and may be considered as representative of the body of an occipital vertebra. Nonunion of the odontoid with the body of the second cervical vertebra is readily distinguished by the line of demarcation at its base and the occasional displacement or altered configuration of the dens.

Among the rarer congenital anomalies in this area are absence of the posterior or anterior arch of the atlas or absence of the sides of the posterior arch of this structure, with persistence of the medial portion.

BASILAR INVAGINATION, BASILAR IMPRESSION AND PLATYBASIA.

These terms are used synonymously so frequently that it has become an almost respectable and acceptable error. Nevertheless, they should be separated and used according to their anatomic counterparts. Platybasia is not basilar impression or basilar invagination. By definition, platybasia indicates a flat, broad, wide base and is used correctly by anthropologists for classifying cranial types. Measurement of the basal angle of the skull is made by drawing a line from the foremost point of the foramen magnum to the center of the sella turcica, and from this point to the root of the nose. This angle normally varies from 115 to 150 degrees, and approaches 180 degrees when platybasia is present.

Basilar invagination refers to an indentation of the upper cervical segments into the base of the skull, as if the weight of the cranium causes a sagging of its base against the rigid cervical spine. This results in an arcuate indentation, the base of the skull assuming a convexity directed caudad.

Simultaneously, and to more variable extent, the tips of the petrous pyramids become directed upwards. The malformation is associated with osteomalacia, rickets, osteogenesis imperfecta, cleidocranial dysostosis, hyperparathyroidism, the lipoidoses, and quite frequently with Paget's disease.

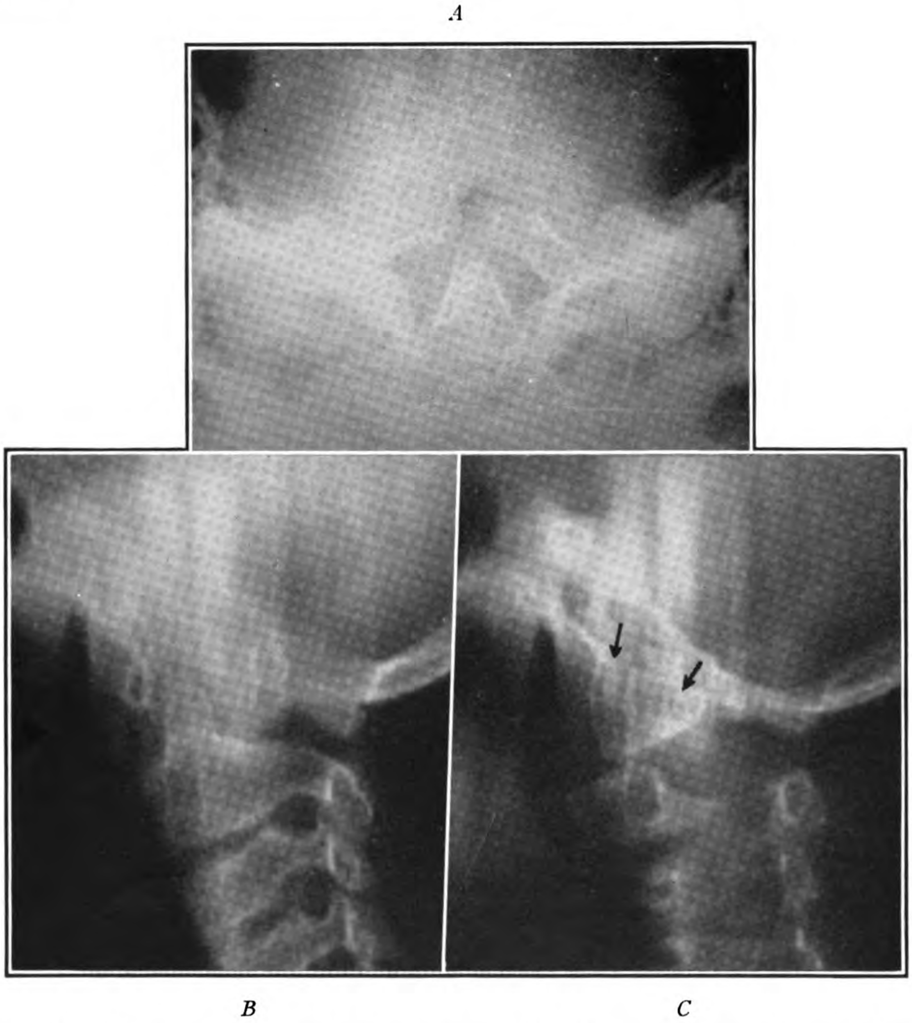

FIG. 58.—*A*, Exaggerated Towne's view showing distorted foramen magnum in patient with basilar invagination. The neural arch of the first cervical vertebra is deformed, its right side being fused to the periphery of the foramen magnum, while the left side is separated. *B*, Same case, midline laminagram. The anterior aspect of the first cervical vertebra is high and fused to the foramen magnum. The first neural arch and spinous process likewise are elevated and impressed into the base of the skull. *C*, Same case, off-center laminagram. The fused body of the first cervical vertebra with the overlying occipital condyle is in focus (arrows).

Deformity of the margins of the foramen magnum and the base of the skull frequently accompanies bony abnormalities of the craniovertebral junction. The foramen magnum becomes misshapen and encroached upon because of assimilation of the atlas or the presence of transitional vertebræ. Because of this the tip of the dens projects into the cervical spinal canal just below and at the foramen magnum, thereby narrowing the space available for the medulla oblongata and the proximal cervical spinal cord.

The incidence of congenital malformation at the occipitocervical junction in patients with basilar impression is frequent. Moreton reported that in a series of 139 cases 41 had associated cervical anomalies. If it were possible to clearly separate those cases of basilar impression based on congenital malformations from those of acquired origin, the figure would probably be higher. The coexistence of other malformations such as fused vertebræ and spinal neural arch deformities should alert the examiner to investigate the cervicooccipital junction. Body section radiographic investigations are of great help for this purpose.

The diagnosis of basilar invagination and basilar impression may be documented further by various linear measurements. Chamberlain postulated that a line drawn on a true lateral skull roentgenogram from the posterior end of the hard palate to the dorsal margin of the foramen magnum in normal individuals would clear the uppermost tip of the dens. McGregor recommended that a line be drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occipital curve on true lateral skull films. The dens should be situated 1.32 mm. ± 2.62 mm. from this line. Bull, Nixon and Pratt recommended an angle determined by the plane of the hard palate and that of the atlas. Angles of 13 degrees or more were regarded as indicating basilar invagination.

Basilar impression or invagination exists without specific symptoms in a rather large proportion of cases. However, when the dens projects sufficiently into the cervical spinal canal, when there is undue mobility of this structure, or when the foramen magnum is sufficiently encroached upon, neurologic changes result either from direct pressure, vascular compression on the adjacent structures or from interference with the cerebrospinal fluid circulation. This may take place regardless of the anatomic cause, and gives rise to perplexing problems simulating syringomyelia, multiple sclerosis or other degenerative demyelinating diseases as well as pictures mimicing tumors of the posterior cranial fossa or the upper cervical spinal canal. Pressure against the cranial nerves or upper cervical nerves may produce paralysis or symptoms referable to cerebellar or medullary compression. In these patients it becomes of the utmost importance to make an accurate diagnosis because surgical intervention can do much to relieve their conditions.

As a rule congenital malformations are not difficult to differentiate from fusions of the cervicooccipital junction such as may result from osteomyelitis, tuberculosis, old fractures, advanced arthritis or echinococcus disease. Charcot's disease in this area is rare.

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Arnold-Chiari Malformation.—The Arnold-Chiari malformation is a deformity of the cerebellum and the brain stem in which these structures project for varying degrees through the foramen magnum into the upper cervical spinal canal. The condition was reported by Arnold (1894) in a description of a newborn infant with a lumbosacral spina bifida together with a large portion of cerebellum extending downward through the foramen magnum in a tongue-like manner overlying the dorsal aspect of the spinal cord. Chiari (1891) delineated three general types divided according to the extent of displacement of the hind-brain into the cervical spinal canal. He pointed out that the cerebellar tonsils and a variable portion of the inferior lobes of the cerebellum may project downward so that the fourth ventricle was not brought below the level of the foramen magnum. The second degree of this change was one in which the lower half of the cerebellum and the adjacent brain stem were brought down into the cervical spinal canal, and the most advanced form was associated with a herniation of practically the entire cerebellum into a cervical spina bifida and meningocele. These early investigators noted a relatively constant association between the malformation and various degrees of meningomyelocele and hydrocephalus. Their observations were chiefly of newborn children and infants, in whom the degree of displacement downward was such that the fourth ventricle was brought into the upper spinal canal.

The neuro-anatomic complications accompanying the various malformations associated with the Arnold-Chiari malformation were described in considerable detail by Lichtenstein (1942). He pointed out that in many instances of spina bifida an abnormal fixation of the spinal cord was present. This prevented its adequate rostral migration with continued development, and the abnormal fixation in some cases resulted in a wide variety of local and distal neuro-anatomic alterations. Included in this list by Lichtenstein were a short cauda equina with a low lying conus medullaris, abnormal stretching of the spinal cord above the site of fixation, elongation of the hind brain with displacement of the medulla oblongata into the vertebral canal together with the choroid plexus of the fourth ventricle and parts of the cerebellum, dysplasia of some of the cerebellar folia, and elongation of the lowermost cranial nerves. Impaction against the walls of the foramen magnum produced compression changes in these structures. In addition, stenosis of the aqueduct of Sylvius was noted, together with internal hydrocephalus secondary to obstruction both at the aqueduct of Sylvius and at the fourth ventricle when this structure was impacted into the foramen magnum. Instances of hydromyelia and syringomyelia-like cavitations in the cervical portion of the spinal cord were also found, and there was evidence of compression on the anterior spinal arteries and the vertebral system of veins at the foramen magnum.

Penfield and Coburn (1938) believed that the Arnold-Chiari malformation resulted from traction on the brain stem in embryonic life resulting from fixation of the spinal cord at a meningocele site. Normally the growth of the vertebral column and the spinal cord are equal until about the third month of uterine life, after which the vertebral canal grows more rapidly than the spinal cord. When spina bifida and meningocele are present the cord may become fixed by the herniation and adhesions at the site of the defect. Traction exerted on the brain stem by the continued growth of the vertebral column pulls the hindbrain down through the foramen magnum. Penfield and Coburn held that this theory was supported by their operative observations that the downward protrusions of the cerebellar tonsils retracted more than 3 cm. on being freed from their attachment to the medullary canal. They also noted, as did Russel and Donald (1935), that the spinal nerve roots in the cervical region were found to pass upward to their intervertebral foramina rather than horizontally, and considered this an additional evidence of downward traction on the cerebrospinal axis.

Barry, Patten and Stewart (1957) regarded the basic cause of the Arnold-Chiari malformation as related to different degrees of overgrowth involving several parts of the neural tube. No need was adduced to postulate traction on the rhombencephalon exerted by means of a structure with as little tensile strength as the embryonic spinal cord. They regarded the hydrocephalus sometimes present as a sequel to the protrusion of the medulla and cerebellum into the spinal canal.

The Arnold-Chiari malformation has often been encountered in association with congenital anomalies at the occipito-cervical junction, such as occipitalized vertebræ and anomalies or failure of fusion of the posterior aspects of the upper cervical segments. Other deformities frequently associated both with the Arnold-Chiari syndrome and meningocele are lacunar skull, congenital and acquired basilar impression and the Klippel-Feil malformation.

The absence of other congenital anomalies of the spinal axis with the Arnold-Chiari malformation has received attention. This has been considered (McConnell and Parker, 1938), as being a factor which tends to cast some doubt on the traction theory of the genesis of the hind-brain malformation. In these cases, at least, some other type of mechanism besides traction would have to be considered as a cause for the cerebellar tonsillar herniation.

Symptomatology associated with the Arnold-Chiari malformation varies greatly. When there is obstruction to the circulation of cerebrospinal fluid because of impingement of the outlets of the fourth ventricle, the outstanding symptoms may be those of hydrocephalus. In others, the clinical picture may be highly suspicious of a cerebellar tumor, with papilledema, vomiting, visual disturbances, and palsies of the cranial nerves. In addition pain may appear in the occipital, sub-occipital or high cervical region due to compression of the upper cervical nerve roots. Not infrequently paresthesias appear in the upper extremities, and it has been noted that this pain sometimes can be relieved by stretching or forward flexion of the neck. These symptoms are not constant, but show a definite periodic tendency. Sometimes they are relieved by rest, only to recur after a sudden sneeze or forced movement which may jam the cerebellar tonsils further into the upper cervical spinal canal. Syrinx-like cavitations in the upper cervical spinal cord may produce a picture much like that of syringomyelia, with dissociation of touch and pain and temperature sensibilities over the affected areas. Not infrequently the disease is confused with other neurologic conditions such as multiple sclerosis or various types of degenerative lesions.

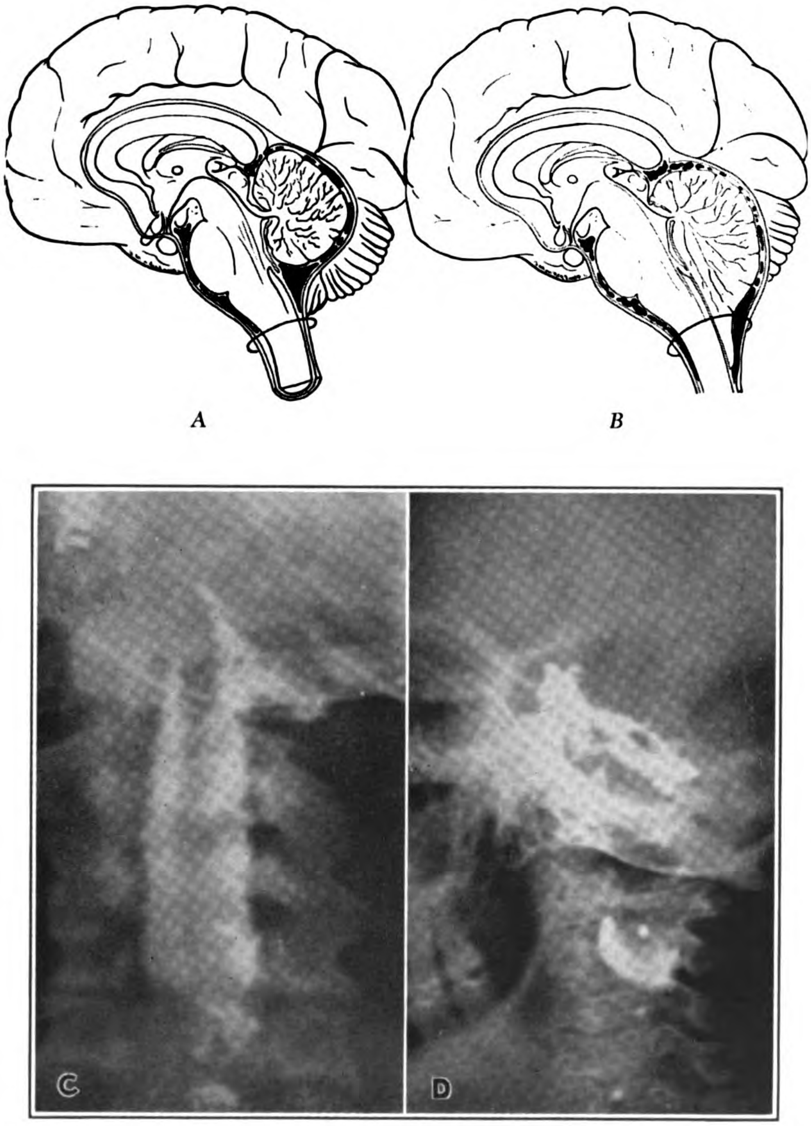

FIG. 59.—*A*, Diagram of normal cisterna magna and cisterna pontis. *B*, Diagram of intrusion of Arnold-Chiari malformation into the cisterna magna. *C*, Pantopaque in normal cisterna magna. *D*, Deformity of cisterna magna by Arnold-Chiari malformation. The pantopaque column is displaced, and at its upper aspect is a semicircular deformity corresponding with the tips of the cerebellar tonsils in the upper cervical spinal canal. (From Epstein, B. S., Am. J. Roentgenol., *59*, 359, 1948, courtesy of Charles C Thomas, Publisher.)

Myelography provides the most accurate approach to the diagnosis. The characteristic deformity of the pantopaque column is produced by the downward protrusion of the cerebellar tonsils, resulting in lobular filling defects in the upper cervical canal. In those instances in which the defect is bilateral a bilobed appearance results, the two arcuate impressions into the pantopaque column being separated by a vertical central bar. If one cerebellar tonsil is lower than the other the defect may appear on one side only, or may be asymmetrical. In some instances with a pronounced degree of protrusion the defect produced by the herniated tonsils may assume an almost semicircular configuration, with thin lateral columns of pantopaque extending upwards towards the cisterna magna.

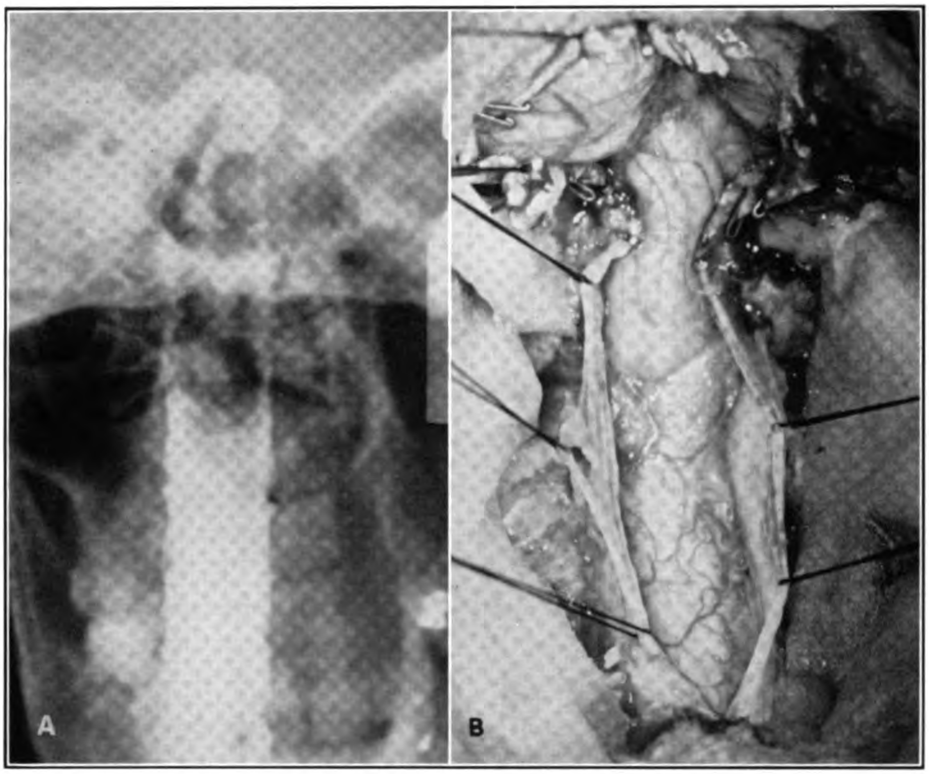

FIG. 60.—*A*, Frontal cervical myelogram showing arcuate deformity of head of pantopaque column at level of second cervical vertebra due to Arnold-Chiari malformation. *B*, Same case as Figure 45 *A*, photograph of Arnold-Chiari malformation *in situ*. (Courtesy of Malis, L. I., Cohen, I., and Gross, S. W., A. M. A. Arch. Surg., *63*, 783, 1951.)

The lower limit of the filling defect may vary considerably. In some cases it may be at the second or third cervical interspace. Advanced downward extension of the cerebellar tonsils as far as the fifth or sixth cervical segment has been described. An unusual example of an atypical filling defect secondary to the Arnold-Chiari malformation was described by Colclough (1940) who reported a case in which the clinical picture simulated that of a herniated cervical disc. According to him, no previous cases had been reported where this developmental abnormality presented a history and symptomatology together with myelographic findings consistent with the diagnosis of herniation of a cervical intervertebral disc. The myelographic defects seen in his cases were at the level of the seventh cervical segment.

Good visualization of the cisterna magna can be obtained myelographically. In the presence of the Arnold-Chiari malformation the usual configuration of this cistern is altered by the impingement of the postero-inferior aspect of the cerebellum as it is drawn downward, producing a curvilinear indentation concave caudally. The arcuate or lobulated border produced by some cerebellar herniations may simulate intradural extramedullary tumors such as neurofibromas or meningiomas. This is more likely to occur when the Arnold-Chiari defects presents a curved configuration due to elimination of the central incisura because of the presence of adhesions. However, if pantopaque is introduced into the cisterna magna the differential diagnosis may be facilitated

Herniations of the cerebellar tonsils also can be identified on gas myelograms, and should be looked for on lumbar pneumoencephalograms. The inferior poles of the herniated tonsils appear as rounded densities which may be large enough to fill the space in and below the foramen magnum.

The diagnosis of the Arnold-Chiari malformation, with or without concomitant skeletal malformations, assumes considerable importance in the presence of symptoms which might be relieved surgically.

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Congenital Absence of the Dens.

This infrequent congenital malformation is due to failure of development of the dens. The lateral masses of the first and the body of the second cervical vertebræ retain their normal relationships. The condition frequently remains symptomless, and is not discovered until radiographic examination either discloses the defect incidentally or after trauma has precipitated symptoms. These vary from vague pain at the base of the skull and upper cervical area to sharp neck pain. Weakness of the upper extremities, tingling, hyperesthesia and paresthesia also have been noted. More advanced pressure effects have resulted in quadriparesis. Bladder and bowel symptoms have not been recorded.

Radiographic examination for congenital absence of the dens is best made using the open-mouth technic and with laminagrams. Being devoid of the usual upward projecting odontoid the medial aspect of the body of the second cervical vertebra assumes a flattened appearance. The relationships of the first and second cervical vertebræ may be disturbed as a result of trauma. In these instances lateral displacement of the atlas as well as rotary posterior displacement can be demonstrated radiologically.

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Congenital Absence of Pedicles and Articular Facets of Cervical Vertebræ.—Among the various congenital anomalies occurring in the cervical spine, the congenital absence of a pedicle is a relatively infrequent occurrence and usually is of little clinical importance except from a diagnostic point of view. The condition may be confused with bony changes incident to malignant disease or possibly neurofibromatosis. The deformity is probably the result of an aberration of segmentation arising in the first eight weeks of fetal life. As a rule the condition is not associated with any specific complaint. Overton and Grossman (1952) pointed out that changes in the planes and angles of articulation of the apophyseal joints may result in instability and may be associated with pain.

From the radiologic viewpoint the diagnosis is most readily made on oblique stereoroentgenograms of the neck. In such cases the involved area presents an unusually large intervertebral foramen with no other evidence of adjacent bony distortion. If the deformity is somewhat more advanced there may be associated changes in the adjacent neural arch, such as absence of the associated lamina or articular processes. Deficiencies in the transverse processes have also been noted in association with this condition. According to Hadley (1952) this deformity may be multiple, so that several pedicles may be absent. Steinback, Boldrey and Sooy (1952) noted that the contralateral lamina in case of congenital absence of the pedicle and superior facet of the seventh cervical vertebra was developed to a greater extent than normal and that this had articulated with the posterior surface of the lamina of the suprajacent sixth cervical vertebra. Due to the increase in size of this lamina and the consequent tilt of the spine, the neural arches of the fifth and seventh cervical vertebræ were in close proximity to each other. In their case the superior articular facet and the posterior portion of the transverse process on the right side of the seventh cervical vertebra were absent, and the anterior or costal portion of the transverse process remained.

Recently I saw a one-year-old child who had a small occipital meningocele. This youngster had congenital absence of the neural arches of the first and second cervical vertebra as well as an extension in the posterior aspect of the foramen magnum through which the meningocele protruded. No neurological disturbances were noted. The bodies of the second and third cervical vertebræ were fused.

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Klippel-Feil Deformity.—This structural deformity of the spinal column originates in intrauterine life. It is of unknown etiology, and usually involves the cervical vertebræ. While the abnormalities described in conjunction with this term vary greatly, the malformation is principally one of fusion of two or more cervical segments. The numerical variations may be great. Varying degrees of alterations occur in the configuration of the vertebræ, from partial fusion of two elements to complete fusion of the entire cervical spine into one solid bony mass. The number of cervical

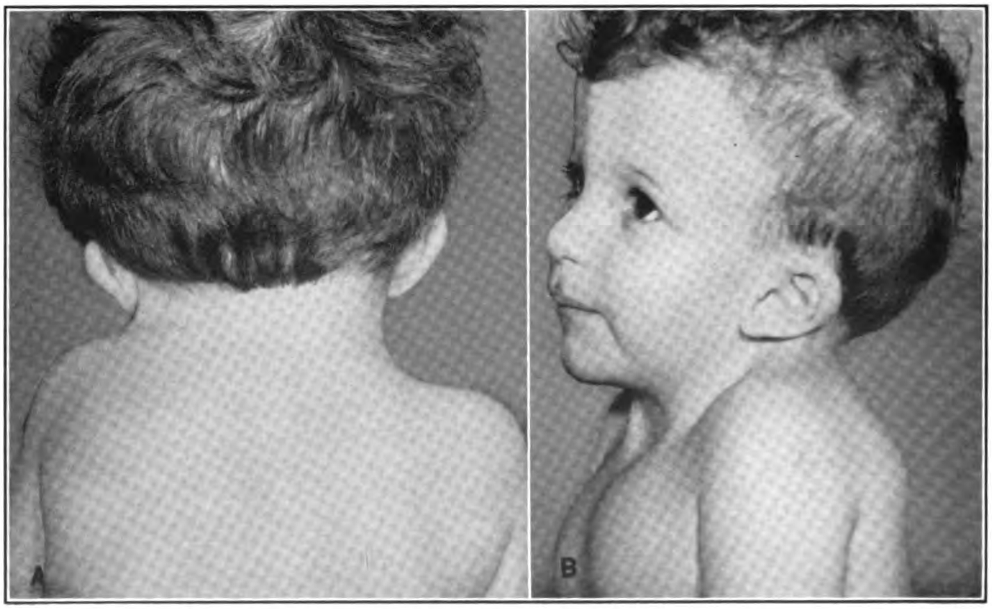

Fig. 61 Legend on opposite page.

segments may be normal or diminished, and there may be concomitant irregularities in the laminæ and spinous processes such as varying degrees of irregular fusion of these structures. Associated with this may be congenital alterations in the upper ribs, cervical ribs, cervico-occipital fusion and various bizarre types of synostosis. The term has also been used to designate a malformation of the spine consisting of a combination of rachis-

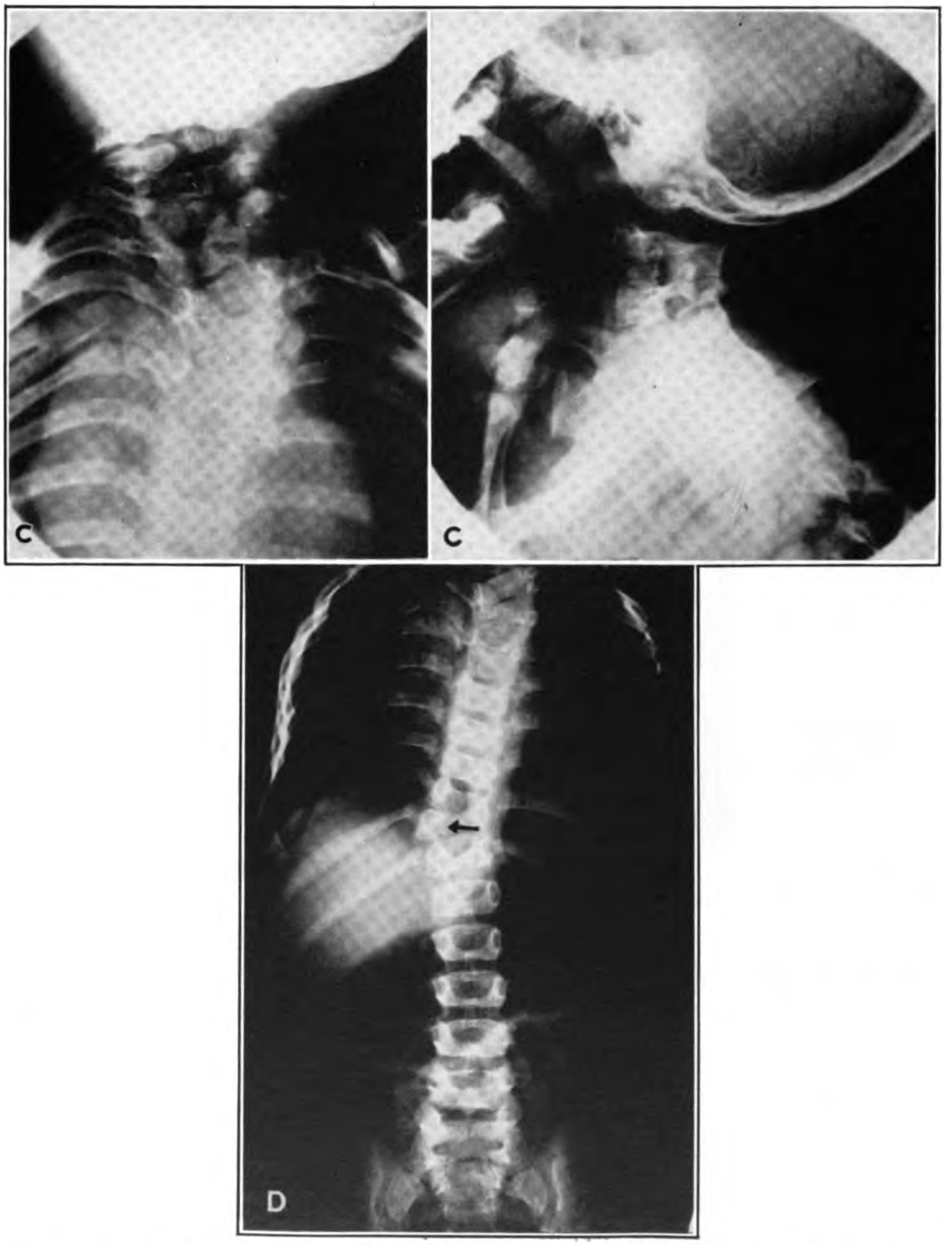

FIG. 61.—*A* and *B*, Photographs of child with Klippel-Feil deformity. Note the short neck and the low hair line. *C*, Same case. The cervical vertebræ are deformed and deficient in number. The upper thoracic vertebræ likewise are malformed. *D*, Same case. The thoracolumbar spine shows a curvature towards the right. Note the hemivertebræ in the lower dorsal region (arrow).

chisis, hemispondylia, block vertebræ and clefts of the neural arches. The involved portion of the spine, may be shortened, kyphotic or scoliotic. The sites of predilection for these changes are the cervicodorsal and dorsolumbar regions.

Considering the Klippel-Feil syndrome as a deformity principally of the cervical spine, a rather typical clinical picture has been worked out. The condition is characterized by shortness of the neck and lowering of the hairline together with limitation of movement of the head. Other abnormalities which sometimes occur include torticollis, facial asymmetry, a lowered nipple line, kyphoscoliosis, and occasionally hemivertebræ. When there is merely synostosis of a couple of cervical segments, very little in the way of clinical symptoms follows. In the more marked cases difficulty in swallowing may appear, and in some neurologic manifestations of radiculitis of various cervical nerve roots, basilar impression defects of the foramen magnum and syringomyelia have been noted. Coexistence of Sprengel's deformity with the Klippel-Feil syndrome has been reported by Furst and Ostrum (1942) and Erskine (1946). In this situation a high position or incomplete descent of the scapulæ is associated with the vertebral anomaly. Facial asymmetry, absence of the external auditory meati, deafness and mental deficiency may also be present.

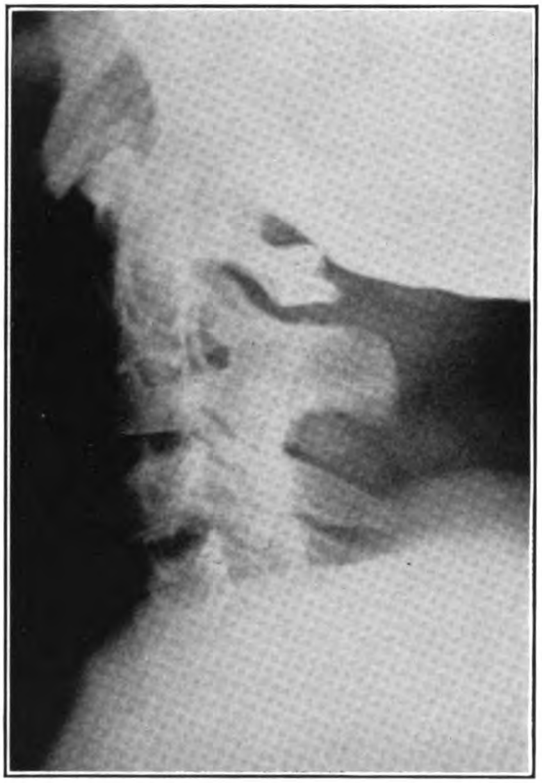

FIG. 62.—Klippel-Feil deformity. The neural arches and vertebral bodies of the second, third and fourth cervical vertebræ are fused.

The more distorted cervical vertebræ can affect the blood supply to the head, particularly in the vicinity of the vertebral arteries. Sudden changes in the position of the head may cause sufficient transitory change to cause loss of consciousness. The association of the Klippel-Feil malformation with central nervous system disorders indistinguishable from syringomyelia or other degenerative conditions has been commented on, and it has been suggested that the disorder may involve the neurogenic as well as the skeletal structures.

Myelographic investigations in patients with the Klippel-Feil syndrome have disclosed no specific disturbances. In those in whom only block vertebræ were present the flow of pantopaque was normal. In others, in whom considerable deformity of the cervical vertebræ existed the caliber of the canal reflected the bony changes, and ridge-like defects were observed.

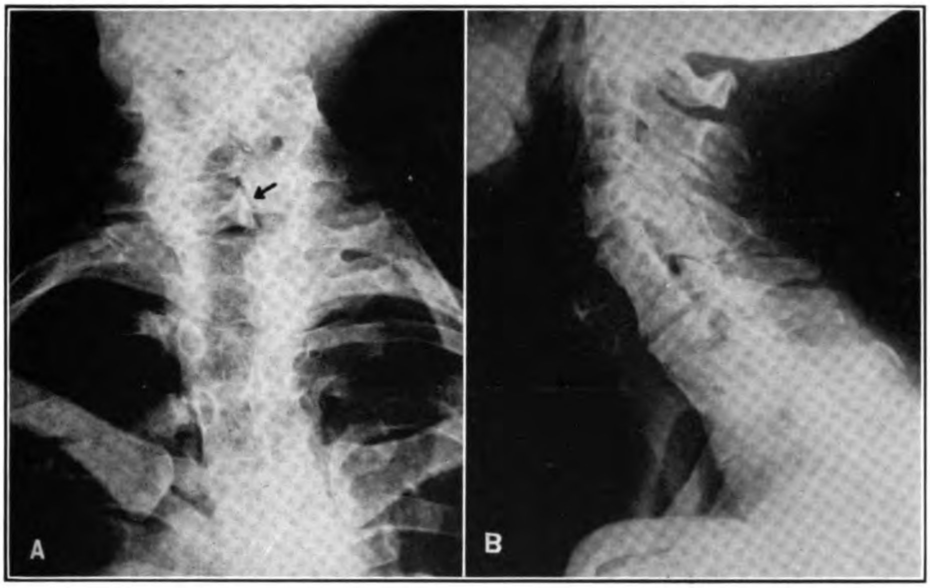

FIG. 63.—*A*, Klippel-Feil deformity. Spina bifida of lower cervical vertebral arches (arrow). *B*, Same case, lateral view. The bodies of the fifth and sixth cervical vertebræ are fused. The seventh cervical vertebra is partly fused to the first thoracic vertebra, which is fused to the one beneath.

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Block Vertebra.—This condition, sometimes referred to as congenital synostosis, involves principally the lumbar vertebræ. The lesion consists of a partial or complete fusion of two or more lumbar segments. This change follows a derangement of embryologic development of the segmental scleromeres, and may be on the basis of complete chondrification of the

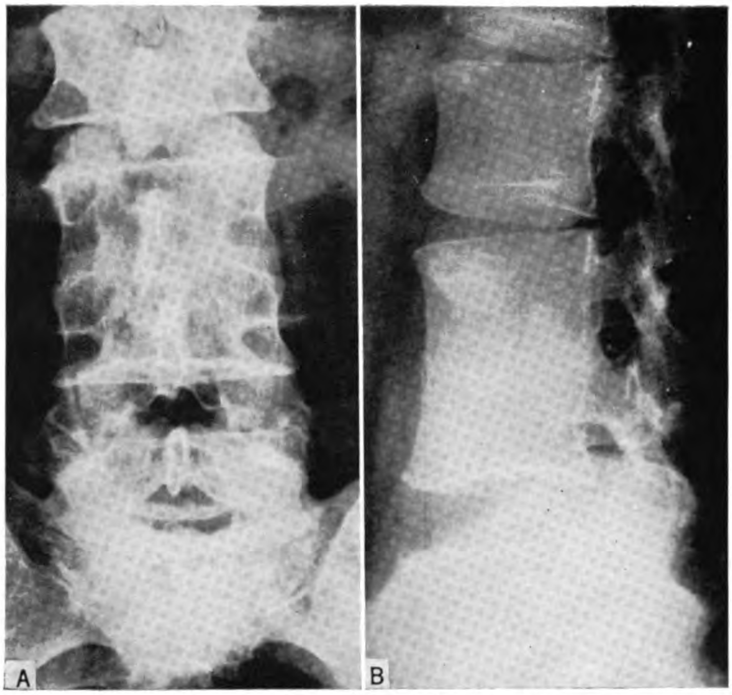

FIG. 64.—*A*, Complete fusion of third and fourth lumbar vertebræ. The height of the vertebral bodies is not grossly changed, but the intervertebral disc is united. *B*, Same patient, lateral view. The area of the intervertebral disc is represented by only a faintly thickened line. Note the diminution in the length of the intervertebral foramen as compared with the one above and below. The neural arches are not fused. The lordotic curve is straightened.

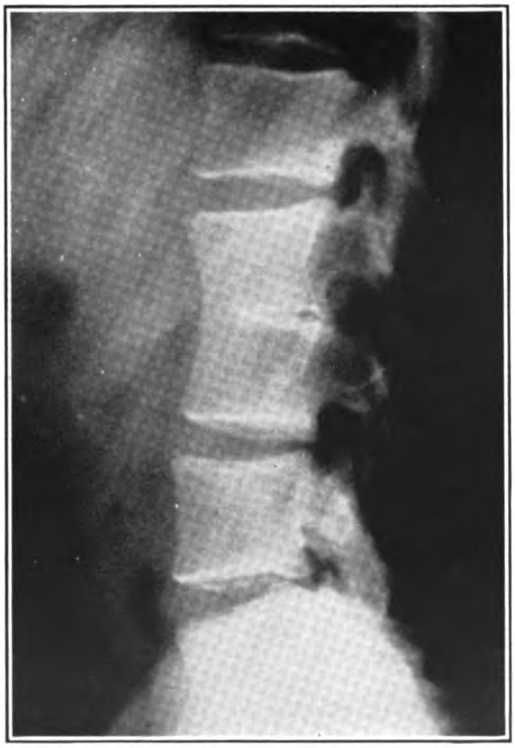

FIG. 65.—Lateral view of incompletely fused second and third lumbar vertebræ. A portion of the dorsal aspect of the intervertebral disc is still present.

dense mesenchymal zone which normally forms the annulus fibrosus. The cartilaginous bodies of the pre-osseous vertebræ become fused and ossify into a solid mass after the periosteal vessels penetrate into the cartilage.

This condition often is identified in the course of x-ray examinations for other conditions. The presence of block vertebræ is not necessarily accompanied by clinical symptoms. Fusion of the vertebral bodies usually produces a fairly straight configuration. Remnants of the intervertebral discs persist occasionally between the fused segments. These cartilaginous streaks may pass from the dorsal aspect of the involved vertebral bodies practically to its anterior border. In cases in which the fusion is relatively complete no such line of demarcation can be observed. The intervertebral formania between block vertebræ often are considerably foreshortened, and the pedicles of the involved vertebræ decreased in size. The neural arches may approach each other, and fuse. The spinous processes may be partially or even completely fused, and represented as a solid block of bone.

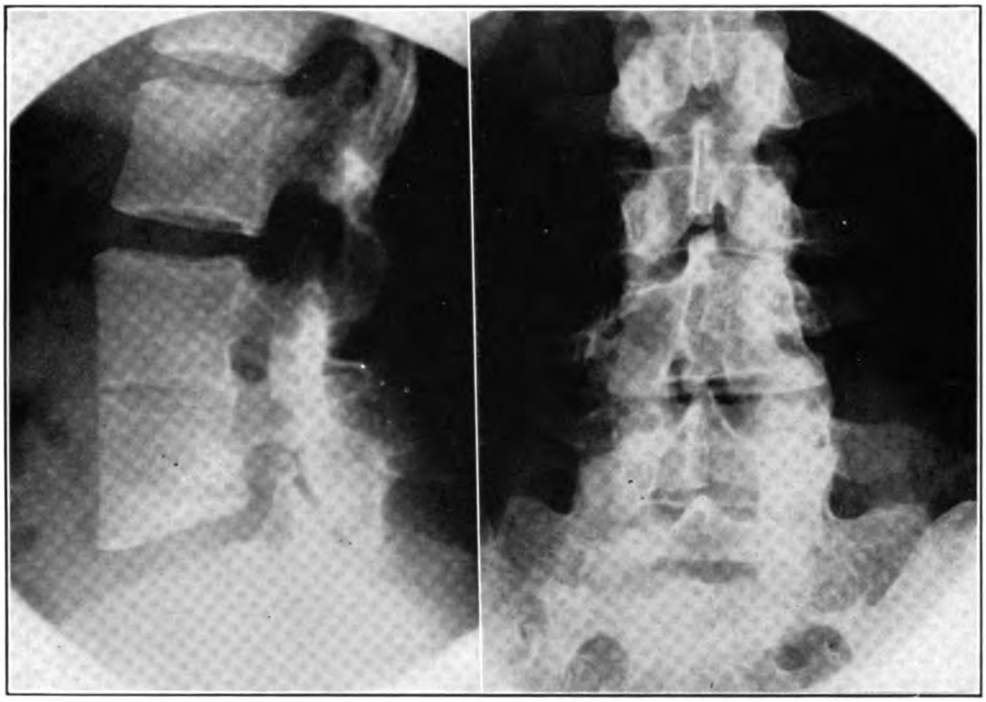
A *B*

FIG. 66.—*A*, Lateral view showing partly fused third and fourth lumbar vertebral bodies. Note the increased height of the fourth vertebra particularly in its anterior aspect. Some intervertebral disc remains. The intervertebral foramen is diminished in size and the neural arches are partly fused. *B*, Anteroposterior view. The left lateral aspect of the fourth lumbar vertebra is higher than its right side. The intervertebral disc is wider on the right than on the left side.

Block vertebræ in the thoracic region are less frequently encountered than in the cervical or lumbar spine. Such deformities in the thoracic spinal column are frequently associated with congenital anomalies of the ribs in which the posterior aspect of the respective ribs become fused and the concomitant vertebral articulations irregularly placed.

Failure of growth of one of the involved vertebræ in the lateral or anterior aspects also occurs, and in such instances a slight or moderate angulation at the point of deficient growth takes place. The heights of congenital partially or completely fused vertebræ are of approximately proper proportions by measurement in some instances, while in others an increase of from 10 to 20 per cent in height is present, the predominant increase occurring ante-

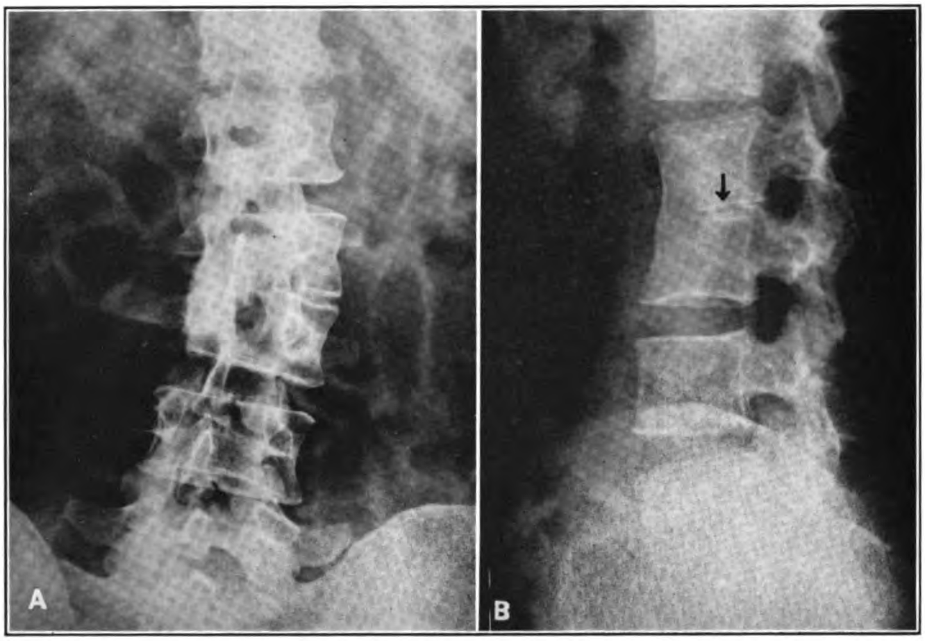

FIG. 67.—*A*, Anteroposterior view showing incomplete fusion of second and third lumbavertebræ, with lateral curvature convex towards the left. Note the persistent intervertebral disc on the left side, while the right is fused. *B*, Same patient, lateral view. The remnant of interr vertebral disc is seen at the posterior aspect of the interspace. The lumbar curve is straightened.

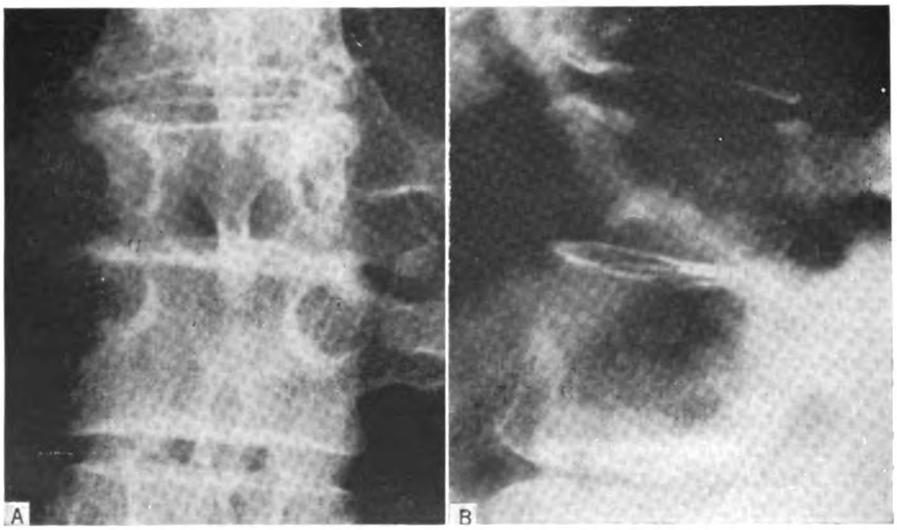

FIG. 68.—Fused ninth and tenth thoracic vertebræ. The persistent intervertebral disc is symmetric. Note the disparity in the heights of the vertebræ, the tenth being considerably higher than the ninth.

riorly. When asymmetric fusion exists varying changes in the alignment of the vertebral bodies results in straightening of the usual spinal curves or tilting towards the side of diminished height. Some authorities consider block vertebræ a variant of the Klippel-Feil syndrome.

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Congenital Lumbar Ribs.—This unusual and clinically unimportant condition sometimes is identified on lumbar spine roentgenograms as well as on films taken in the course of investigations which frequently are unrelated to the vertebral column. They occur more often in females. Lumbar ribs usually present as symmetrical structures which vary in length from a few millimeters to 5 or 6 centimeters. Instances in which these anomalous ribs are asymmetrical also have been described. Lumbar ribs usually present well-formed articulations with the transverse processes of the first lumbar vertebra. The osseous structure of the rib is normal bone undistinguishable from those above. Care must be taken in the diagnosis of a lumbar rib to make sure that there is no anomaly of the fifth lumbar vertebra which would lead to a false count. If there is any question as to a lumbar rib being present, a roentgenogram of the entire spine might be required before the diagnosis can be established. The length of the suspected lumbar rib offers no definite clue as to its presence, inasmuch as a normal twelfth rib might be short but otherwise unremarkable.

Sacral ribs are rare. Infrequently tubular, elongated ossicles form in the lateral aspect of the sacrococcygeal area, situated on either side between the first coccygeal and the last sacral vertebræ.

Cervical Ribs.—This frequent and sometimes clinically important anomaly represents a progressive development of the embryonic costal processes of the seventh cervical vertebra. The remainder of the cervical spine does not show similar maldevelopment. The radiologic diagnosis of cervical ribs is dependent upon the demonstration of a costal vertebral joint. Cervical ribs may develop to a widely varying degree, and present a wide variety of anomalous configurations. Some sharply terminate a relatively short distance from the joint, others extend as much as 5 to 6 centimeters parallel to the course of the true first rib. Often the cervical ribs fuse distally with the subjacent first ribs. Anomalous configurations of the distal end of the first and sometimes of the second ribs also occur. Asymmetry of cervical ribs is quite common.

In childhood the costal processes remain ununited until some time between the fourth and tenth years. During the earlier years these are independent of the transverse processes, and should not be interpreted as cervical ribs. The developing costal processes, which lie anteriorly, and the posteriorly situated transverse processes later unite to encircle the foramen transversarium. These foramina are usually not seen on cervical roentgenograms in adults, but may be identified occasionally in children. When cervical ribs exist, they are present at birth. It may be difficult to identify them from normal costal processes, but if their anterior and downward curvatures follow the course of the true ribs one may arrive at a reasonably correct conclusion.

In the x-ray demonstration of cervical ribs it has been found helpful to tilt the tube towards the head about 10 to 15 degrees in an effort to delineate the inferior margin of the suspected cervical rib from the adjacent first rib. In addition to this overpenetrated and oblique roentgenograms sometimes help identify an articular space which otherwise might be hidden.

Another anomaly of the seventh cervical transverse processes which at times may assume an importance much the same as cervical ribs is a prolongation of the transverse processes.

While cervical ribs are frequently asymptomatic, and often are identified on routine roentgenograms of the chest, the condition nevertheless may be associated with clinical symptoms, notably pain referable to the neck, shoulder or supraclavicular region. This may be accompanied with severe pain extending down the arm following the course of the compressed segments of the brachial plexus. The pain varies considerably in intensity, sometimes being sharp, at other times being of a dull, aching nature. Sensory disturbances may be associated with the pain, and if the lesion is such that the vascular structures of the base of the neck are embarrassed, changes referable to such pathologic alteration of the circulation may appear. Surgical intervention is sometimes required for relief of symptoms in severe cases.

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Developmental Anomalies of the Vertebral Bodies.—In order to understand congenital deformities of the vertebral bodies one must keep in mind their embryologic development from the notochordal to the mesenchymal state, thence into cartilage and finally differentiation into bone and intervertebral discs. Aberrations in growth occur at any time during the course of development, and the consequent deformities bear a relationship to the stage of growth when this supervened. The important fact that alterations in blood supply influences the pattern of growth likewise must be remembered. Ossification depends on the vascular supply, and if either the dorsal or ventral distribution to the respective ossification centers fails to appear there follows maldevelopment and failure of growth in that area. If there is vascular aplasia, the vertebral body fails to ossify, and a gibbus formation follows. If the neural arches continue to grow, fusion sometimes takes place anterior to the spinal canal resulting in a small dorsal hemivertebra.

Persistence of the notochordal canal infrequently results in a slender cylindrical deformity best identified on lateral roentgenograms. When this persists and is associated with coronal cleft vertebræ it is characterized by a tubular or spindle-shaped radiolucency in one or more vertebræ. The unossified area contains cartilage and some notochordal remnants. This sagittal tubular defect sometimes disappears in two to four months, and is regarded as a delay in the normal process of recession of the notochord.

Indentations into the superior and inferior surfaces of normal vertebræ in young people indicate pressure against these areas by the bulging nucleus pulposus, and indicate the site of the notochordal recession into the intervertebral disc.

From a radiological viewpoint anomalies of the vertebral bodies may be classified in several groups. The minor malformations frequently have no significance other than the inference that where one congenital variation occurs others should be suspected. The more advanced deformities result in structural alterations, while the most grotesque ones deform the vertebral column so much that recognition of the various segments becomes impossible, and incompatibility with life is not infrequent. The various groups of deformities, which depend mainly on abnormal patterns of ossification and vascularization of the growth centers include the fairly frequent sagittal cleft vertebræ, wedged hemivertebræ in which a lateral half of the body fails to develop, and the less frequently observed dorsal hemivertebræ. Coronal cleft vertebræ are infrequent, and ventral hemivertebræ are virtually unknown.

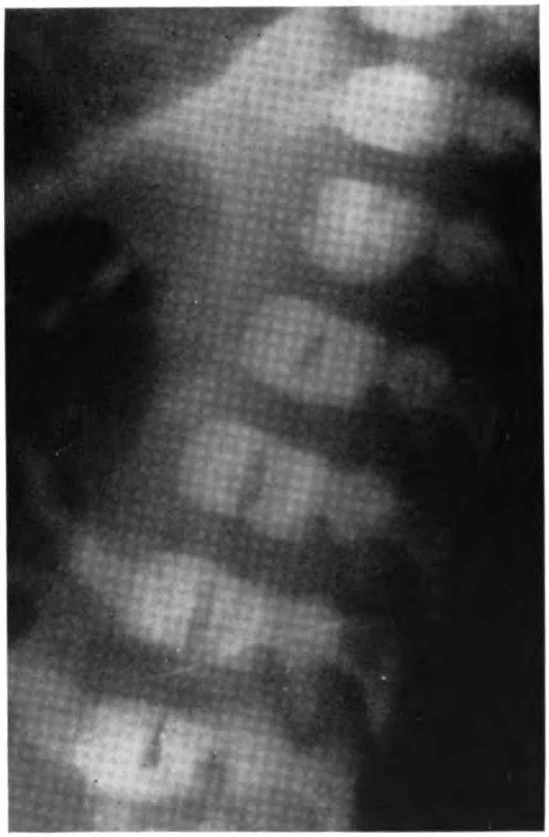

FIG. 69.—Coronal cleft vertebral defects in lumbar vertebræ of a newborn infant.

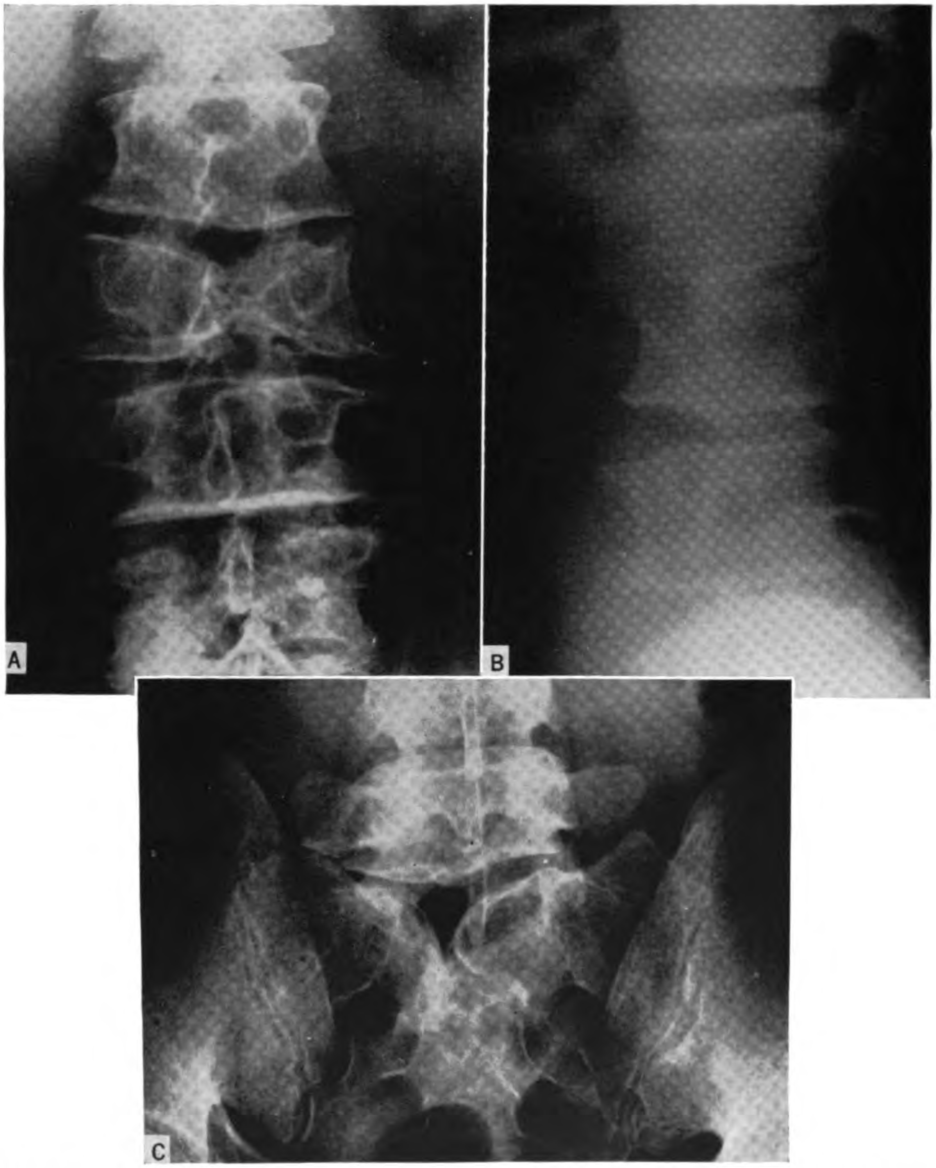

FIG. 70.—*A*, Anteroposterior view, "butterfly" second lumbar vertebra. The right half presents a rounded medial angle, while the left half is more pointed medially. Note the compensatory changes in the vertebræ above and beneath. *B*, Same patient, lateral view. The anterior aspect of the involved vertebra is diminished in height, indicating probable failure of growth of the anterior nucleus on the left side. The sagittal cleft in this instance is somewhat atypical because of this. *C*, Symmetric sagittaly cleft first sacral vertebra.

The concurrence of vertebral malformations with enteric cysts or duplication of the foregut is recognized as a situation dependent on failure of proper notochordal and entodermal development. Veeneklass suggested that if the separation of the notochord from the entoderm is not complete, a pouch of the latter could be withdrawn from the primitive alimentary tract into the mesodermal anlage of the vertebral bodies. Such a pouch could then grow to form a mediastinal or intestinal duplication which may or may not communicate with the alimentary tract. The column of cells attaching the pouch to the notochord may interfere with proper growth of the vertebral bodies or cause failure in fusion of the neural arches. The combination of cleft or otherwise malformed vertebræ with a mediastinal mass should suggest the existence of a neurenteric cyst.

Sagittal cleft vertebræ are attributed to persistence of the ventrodorsal extension of the perichordal sheath, or to failure of fusion of the lateral halves of the vertebral bodies caused by persistence of the notochord. The lateral halves may ossify separately, resulting in the production of hemivertebræ identified as such by the asymmetric development of the ossification center which retains its normal potential. It is unknown whether this represents a failure of vascular supply, to some defect in the cartilage anlage, or some other deficiency in the process of ossification. As development of the normally growing side procedes, it changes from a somewhat cuboidal to a rather triangular configuration of varying proportions and usually with rounded angles. A persistence of a thin radioopaque streak in the

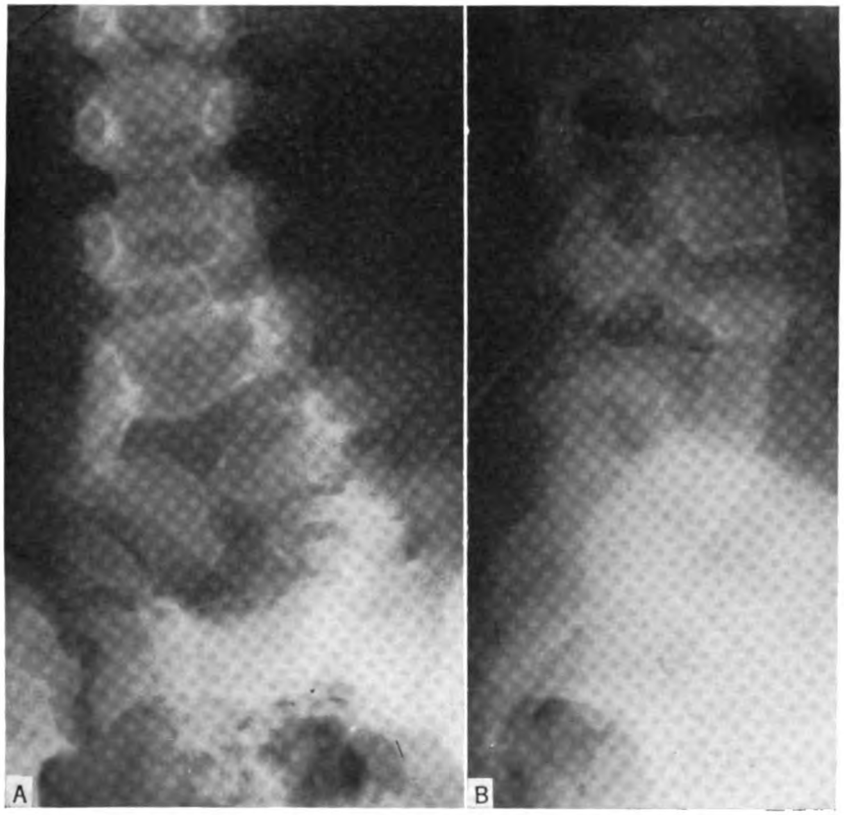

FIG. 71.—*A*, Sagittally cleft and malformed fourth and fifth lumbar and first sacral vertebræ. The lateral halves are asymmetric, and the neural arches are open. The pedicles are malformed and widely separated. *B*, Same case, lateral view. Note the malformation of the fourth and fifth lumbar vertebræ particularly in their posterior aspects.

The heights of these vertebræ are diminished. The malformation may be caused by failure of development of the posterior centers of ossification.

vicinity of the maldeveloped segment sometimes can be demonstrated. This process may involve several vertebral bodies, and if there is asymmetry in the development of the respective lateral halves a spinal curvature results. The neural arches of the involved vertebræ are not necessarily affected, and the development of the ribs and transverse processes is not grossly impeded.

With greater malformation, sagittal cleft vertebræ present considerable separation of the lateral halves. The bodies assume the configuration of two triangles whose apices face one another. The ossification of the vertebral bodies is well maintained. The term "butterfly vertebra" is applied to those in which the two halves are pinched off so that they simulate wings protruding from an imaginary butterfly whose body corresponds to the point where the two triangles meet in the midline. Thin strands of bone sometimes are seen traversing this cleft. The halves may be unequal in size, displaced laterally, or wedged anteriorly with consequent scoliosis and kyphosis.The adjacent intervertebral spaces may be narrowed. In instances where this is fairly symmetric the vertebræ above and below present bulges downward and upward respectively, directed towards the cleft body. The pedicles of the involved vertebra may become rather widely separated, following the increased width of the affected vertebral body.

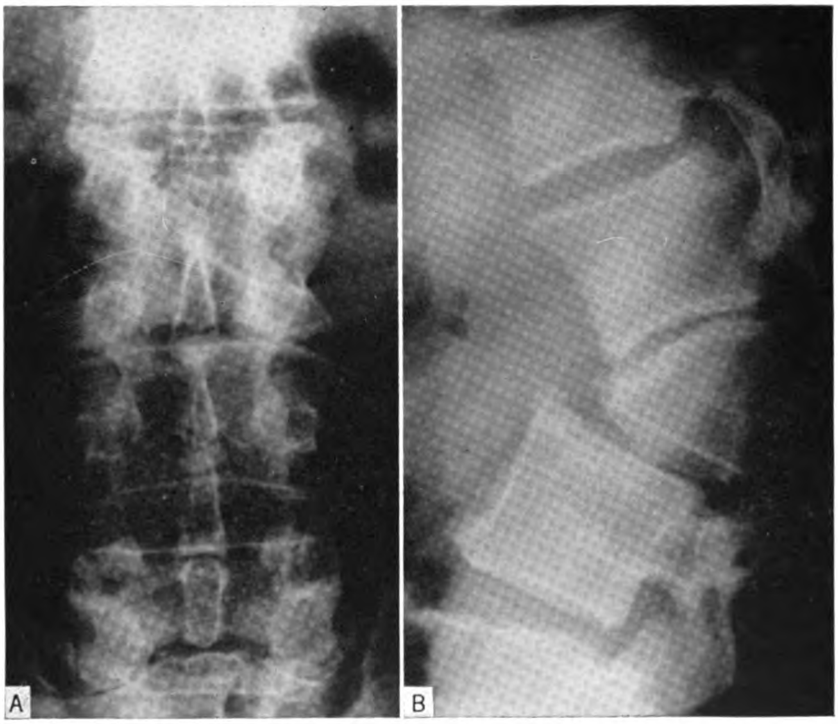

FIG. 72.—*A* and *B*, Wedge Vertebra, with failure of development of anterior nucleus. The dorsal hemivertebra produces a gibbus deformity.

Cave regards the term "butterfly vertebra" as applicable to two situations. The first is the basic form of sagittal cleft vertebra, which presents an appearance designated as the "double D" type, with rounded medial margins. The second, referred to as the "double wedge" form, is attributed to a partial dorsal hemivertebra together with a sagittal cleft. In this form the two halves are more triangular, with sharply formed angles meeting in the midline as the apices of the respective triangles.

Coronal cleft vertebræ in infants results from persistence of the anterior and posterior ossification centers of the thoracic and lumbar vertebræ beyond approximately the sixteenth week of intrauterine life. These clefts are seen more often in the lumbar spine, and affect several segments. A linear or oval deformity with well defined margin is identified, which continued to normal consolidation in a few months. The intervertebral disc is not deformed in these cases.

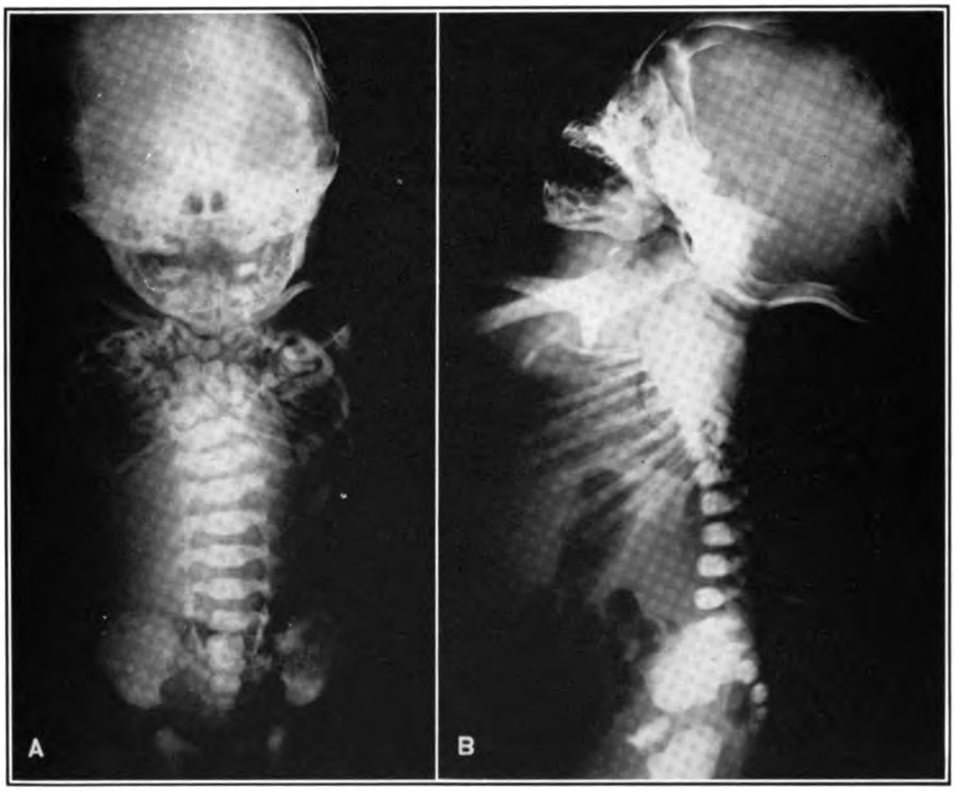

FIG. 73.—*A*, Anteroposterior and *B*, lateral views of multiple congenitally malformed dorsal vertebræ. The architecture has been markedly distorted, so that the individual vertebræ and ribs cannot be identified. Note the lacunar skull and the widened lumbar spinal canal, (meningocele.)

The term "wedged" vertebra refers to that condition in which only a single half of a vertebral body is present, a true hemivertebra. This is attributed to a defect in ossification of the cartilaginous body in early fetal life, and has been ascribed by Junghanns to a local deficiency in blood supply in the affected half. At first the developing half maintains a relatively cuboidal configuration, but with further growth and strain the change in pressure results in a triangular, wedge-like configuration with the apex directed medially. The term also may be applied to some of the instances of sagittal cleft vertebræ mentioned above, particularly those with advanced and multiple malformations.

The ultimate picture of disruption is one in which there is neither pattern nor organization in the distribution of the respective vertebræ. The appearance is one aptly described as a "mixed-up spine," composed of rounded, triangular, cuboidal and sometimes multiangulated structures which seemingly fit into each other to form a vertebral column whose components often cannot be distinguished one from another. In these the disruption often involves the neural arches and pedicles as well, together with malformations of the adjacent ribs and transverse processes. This type of pronounced change has been ascribed to a faulty bilateral cranial hemimetameric shift of the vertebral segments which occur in the very early stages of the growth of the vertebral column. This shift occurring on one side causes an anlage for half a vertebra to remain at the lower as well as at the upper end of the unequally shifted and mismatched vertebræ. The more advanced forms of this is incompatible with life.

A relatively rare vertebral malformation is the dorsal hemivertebra which results from failure of ossification of the anterior ossification center, probably because of failure of the blood supply to that area. The anterior half is reduced to a fibrous strand, while the dorsal half assumes a triangular configuration with the vertebral bodies above and beneath approaching each other. A gibbus deformity results and considerable spur formation often follows as a reparative effort. The uninvolved adjacent bodies sometimes increase their vertical dimensions.

Ventral hemivertebrae might be anticipated when the blood supply to the posterior half of the developing body fails. However, there is insufficient evidence to warrant further comment on this intriguing but apparently very rare possibility.

Complete agenesis of the vertebral bodies may be observed in stillborn infants with marked congenital malformations, and as such have considerable academic but little practical radiologic interest. However, agenesis of the vertebral bodies as a cause of dwarfism has been mentioned by Giannini, Borrelli and Greenberg (1948) who reported a four-and-one-half-year-old male with multiple developmental defects including incomplete development of the thoracic vertebral bodies with a complete lack of several segments. They found that this deformity may not be a true absence of a vertebral body as one understands the term from a literal translation, but may represent a stage or a form of anomalous development of many of the vertebral bodies corresponding to the complete disruption mentioned above.

A considerable number of clinical implications may be drawn from the presence of anomalies of the vertebræ. When the defect is mild the only change of clinical importance may be a relatively fixed and asymptomatic curvature of the spine. However, it may cause some question in differential diagnosis because of some resemblance to tuberculous spondylitis or fracture. If there is a more pronounced degree of deformity, mild or severe pain may be present. Of more interest from a diagnostic point of view is the fact that the existence of even a mild congenital anomaly in the spine may point toward the co-existence of other visceral congenital anomalies of considerable significance.

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Cleft Spinous Processes.—This is one of the common anomalies of vertebral structures and very often is unassociated with any specific clinical complaints. Its mildest form is a lack of fusion of the neural arches of one or several vertebræ. The most common location is in the fifth lumbar neural arch, and the first sacral segment is next in order of frequency. The coexistence of both lesions is not uncommon. In addition one may often observe a variable degree of lack of fusion of the dorsal plate of the lower portion of the sacrum, varying from a small cleft in the distal portion of the bone extending over one or two segments to an almost complete lack of fusion represented by a central radiolucent streak extending for the entire length of the sacrum.

Statistics as to the incidence of failure of fusion of the upper sacrum and the fifth lumbar neural arch vary. Southworth and Bersack (1950) reported that out of 550 patients with anomalies of the lumbosacral vertebræ most had little or no symptoms referable to the low back. Spina bifida occulta was present in 18.2 per cent; 2.2. per cent of these occurred in the lumbar spine and 16.0 per cent involved the sacrum. They analyzed the defects in the neural arches and found that of the lumbar defects only one was to the left of the mid-line. Of the sacral defects 3 were to the right, 2 to the left of the mid-line, and 1 bilateral. The vast majority of the defects were central in location. Dittrich (1938) reported that 5 per cent of all spines examined roentgenologically presented spina bifida occulta, a figure close to that reported by Breck, Hillsman and Basom (1944), who reported an incidence of 6 per cent in a series of 450 cases. However, Friedman, Fischer and Van Demark (1946) reported an incidence of 36 cases of spina bifida occulta in a review of the cases of 100 soldiers. My own experience coincides with the last authors' insofar as frequency of occurrence is concerned. It is not at all infrequent in the course of routine gastrointestinal or genitourinary examinations to encounter a completely unsuspected and symptomless spina bifida occulta of the fifth lumbar vertebra, the first sacral segment or a varying degree of spina bifida of the lower dorsal aspect of the sacrum. Combinations of all of these three are frequent, and together with them the incidence of atypical formation of the articular processes between the fourth and fifth and fifth lumbar and first sacral segments is by no means uncommon.

In addition to cleft spinous processes in the regions named above I have seen frequent instances of lack of fusion of the spinous processes of the

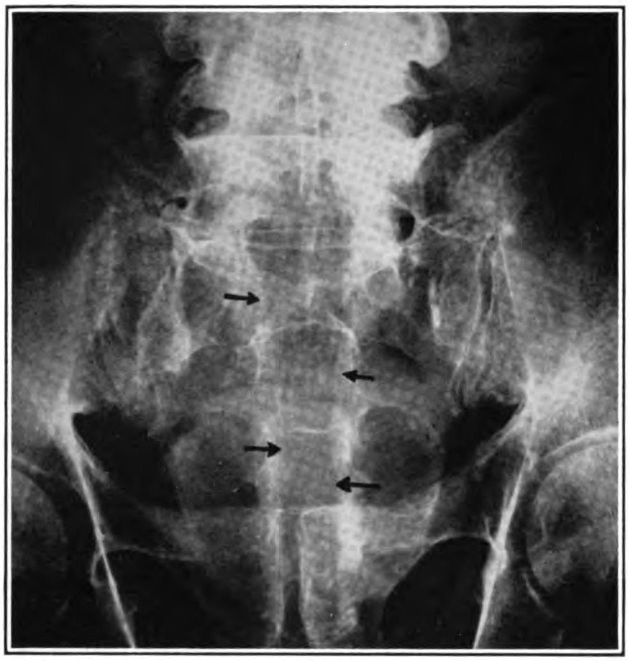

FIG. 74.—Failure of fusion of the dorsal plate of the sacrum extending from the first to the last sacral segment (arrows).

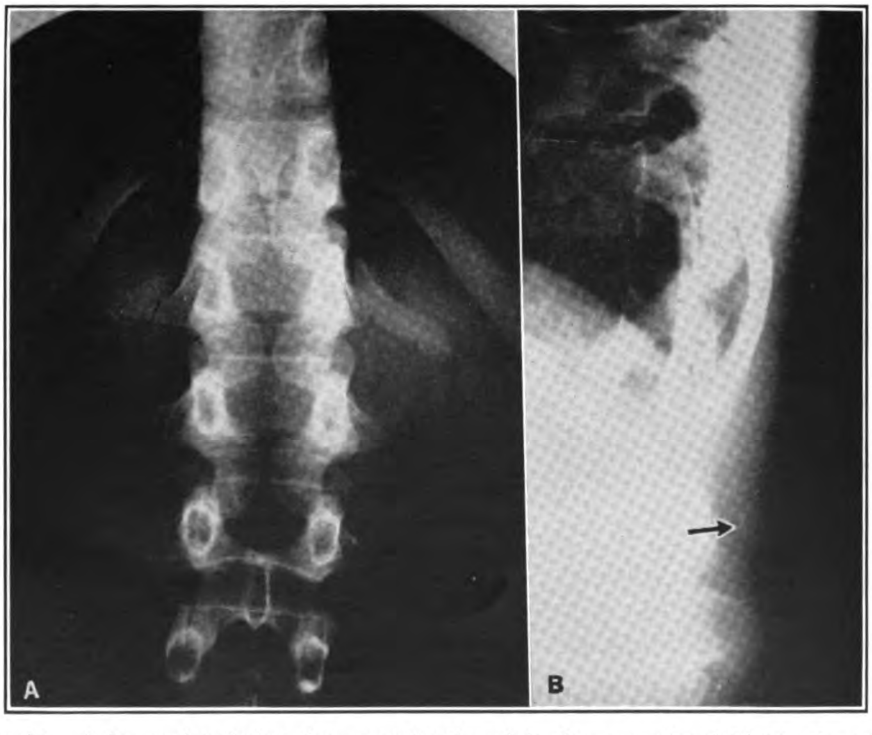

FIG. 75.—*A*, Incomplete fusion of the neural arches of the first, second and third lumbar vertebræ, with absence of spinous processes. Bilateral lumbar ribs. *B*, Same case, lateral view. The spinous processes are missing. A rudimentary ossicle is seen above the normally developed spinous process of the third lumbar vertebra (arrow).

twelfth dorsal vertebra, the first lumbar vertebra, and an occasional case of lack of fusion of the spinous processes of the second lumbar vertebra. Similar changes have also been occasionally demonstrated in the spinous processes of the first and seventh cervical and first and second thoracic vertebræ. These less frequent examples portray the embryologic change more faithfully in that the lesion is represented by a thin radiolucent cleft in the vicinity of the spinous process of the affected vertebral segment. The range of variation in the appearance of the laminæ and pedicles of these vertebræ is less than in the fifth lumbar or first sacral segments. Cleft spinous processes in these regions follow the pattern of fusion of the laminæ in the midline to form the spinous process. It should be recalled that the laminæ fuse at about the first year of life and their development is completed during adolescence. The spinous process is capped at about this time by an apophysis which calcifies completely and fuses with the main mass of the spinous process at about the twenty-fourth or twenty-fifth year of life. This additional apophysis sometimes fails to unite with the spinous process, and the structure may appear as a rather oval calcific shadow overlying the defect in the involved neural arch. This too may sometimes assume an anomalous appearance and may grow to a fairly large hook-shaped bone.

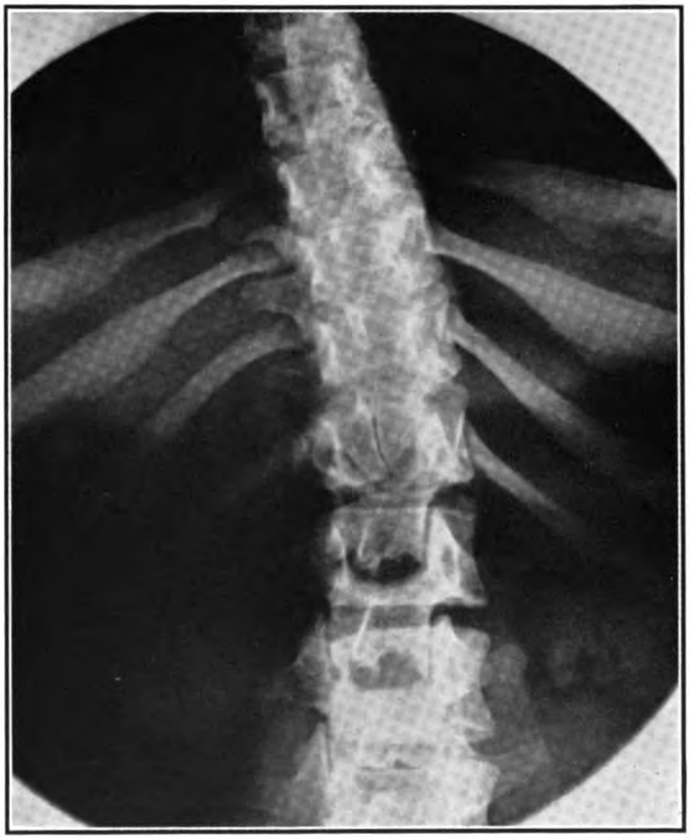

FIG. 76.—Incomplete fusion of spinous process of twelfth dorsal vertebra, with unequal height of that vertebral body. Note the convexity of the spine towards the left.

An instance of this rather rare anomaly associated with considerable back pain is shown in Figure 77.

While most patients with lumbar cleft spinous processes usually have no specific complaints there are certain cases in which the anomaly may be accompanied by low back pain. Such cases are very difficult to evaluate and the defect should not be held as the responsible factor until all other possibilities have been excluded.

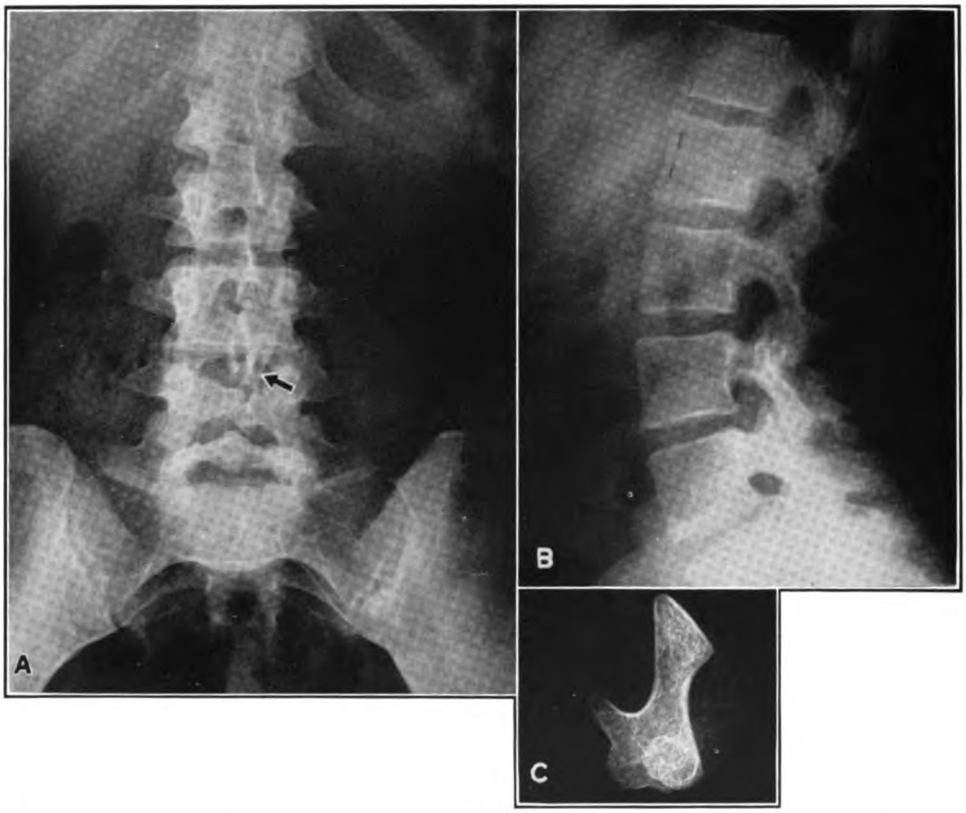

FIG. 77.—*A*, Anteroposterior view of lumbar spine. Incomplete fusion of the spinous process of the fourth lumbar vertebra. Just above this cleft is a linear ossicle (arrow) directed towards the left of the midline. *B*, Same case, lateral view. The ossicle can be seen just above the base of the spinous process of the fourth lumbar vertebra. *C*, Same case. Operative specimen. The ossicle was anchored to the dura by firm adhesive bands.

A very interesting series of cases was published by Jelsma and Ploetner (1953), who reported a group of 18 patients with low back pain radiating to the hips and sometimes to both lower extremities or to one leg. While this pain was sometimes recurrent, it often was continuous over a period of years and could be aggravated by bending. Usually no reflex or sensory changes were noted. Operation disclosed a mass of fibrous or fibrolipomatous tissue, often with a constricting mass of fibrous tissue involving the dura. At times fibrosis involving the sheaths of the emerging nerve roots was encountered. They did not observe cord involvement by traction on the filum terminale. None of their cases showed any evidence of myelodysplasia, and the bony defects were minimal. Apparently the pain was produced by a traction mechanism in which the dense fibrous tissue extended from the dura to an insertion where the posterior spinous processes ordinarily would have appeared, and there the fibrous bands become attached to the lumbar fascia of the back.

Radiologically, lumbar cleft spinous processes are often identified on routine anteroposterior roentgenograms of the lumbosacral spine or abdomen. In certain instances a tilt-up study of the lumbosacral articulation is helpful in identifying the neural arches of the fifth lumbar and first sacral segments.

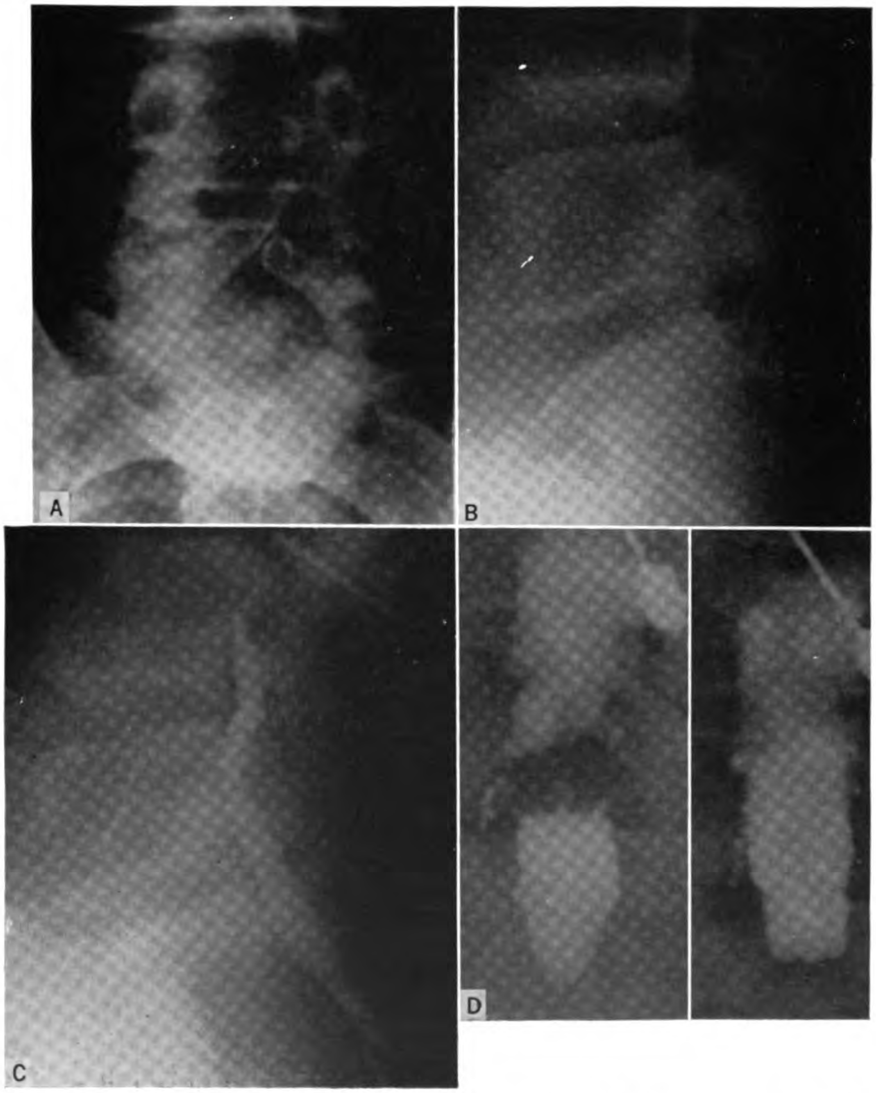

FIG. 78.—*A*, Incomplete fusion of spinous process of fifth lumbar vertebra in a 47 year old man with complaints diagnosed as herniated disc. *B*, Same patient, lateral view. The pedicle of L-5 is heavy, and the lumbar canal appears narrowed sagitally. *C*, Same patient, lateral myelogram. Note the narrowness of the spinal canal and the long caudal sac. At the level of the midportion of L-5 there is an indentation into the column. *D*, The anteroposterior myelograms showed hesitation of flow at the level of the superior aspect of L-5. At operation the defect was found to be due to pressure from a heavy, thickened right lamina, compressing the cauda equina significantly. The discs were normal. Relief followed decompression of the canal.

The configuration of the neural arches in these patients may vary considerably. In some the bifid spinous process may be manifested by a double, rather heavy stump-like shadow of the unfused spinous processes. The cleft may measure from 2 to 5 mm. in width, and its direction may be either sagittal or oblique. In others only one of these calcific shadows may be seen, the other having failed to develop.

The laminæ sometimes develop asymmetrically, and may vary in thickness. The first sacral segment may show a defect which varies from a slight slight downward concavity of the dorsum of the first sacral vertebra to a complete absence of this area. A rather V-shaped deformity of the body of the first sacral segment may co-exist with the spina bifida occulta of its neural arch.

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Diastematomyelia.—This remarkable but infrequent malformation of the spinal cord represents a developmental error associated with disturbance of the growth of the neural plate and the neurenteric canal. In vertebrates the neurenteric canal is a minute connection between the amniotic cavity and the yolk sac. It passes through the primitive node, which migrates caudally as the neural plate evolves into the spinal cord. At no time in its development is the cord duplicated, so that diatematomyelia does not represent a twinning or actual double development of the cord. Persistence of a connection between the dorsal aspect of an ectopic or accessory neurenteric canal is followed by malformations of the cord, the vertebræ and the neural arches. Similar communications between the ventral aspect of the neurenteric canal results in various malformations dependent on the open area. Enterogenous cysts may follow persistence of the midportion, duplication of the bowel from persistence of the ventral portion alone, while if the terminal segments persists, a wide variety of lesions such as cutaneous sinuses, dermoids or teratomas may occur singly or in combination. If both the dorsal and ventral portions of an accessory neurenteric canal persists, various combinations of vertebral and visceral malformations can occur.

The term diastematomyelia indicates the presence of a fissure of the spinal cord. The spur formation often observed in the middle of the defects is attributed to the formation of a central fused pedicle resulting from abnormal growth of the underlying malformed vertebra, which, in the process of growth as a bifid structure, laid down a rudimentary pair of pedicles. When calcified, the central spur is a reliable index to the correct diagnosis. In some instances the spur is cartilaginous, and hence cannot be identified on plain spinal roentgenograms. The bifurcation of the cord, which may extend over several segments, cannot be estimated from the position of the spur.

Diastematomyelia is encountered in both sexes at any time from fetal life to old age. In most of the cases there is a pronounced associated anomaly of the neural arches, and in some meningocele or myelomeningocele may be found. Death from infection of these structures may occur in early life. The lesion occurs anywhere in the dorsal or lumbar spine but appears to be most frequent in the lower dorsal or upper lumbar regions, although instances have been encountered in the mid-dorsal and lower lumbar areas. Splitting of the spinal cord occurs in varying degrees from an abortive doubling with no clinical or radiologic manifestations to division of the cord into parallel structures. In marked cases there may even be doubling of the lower spinal column (Pickles, 1949).

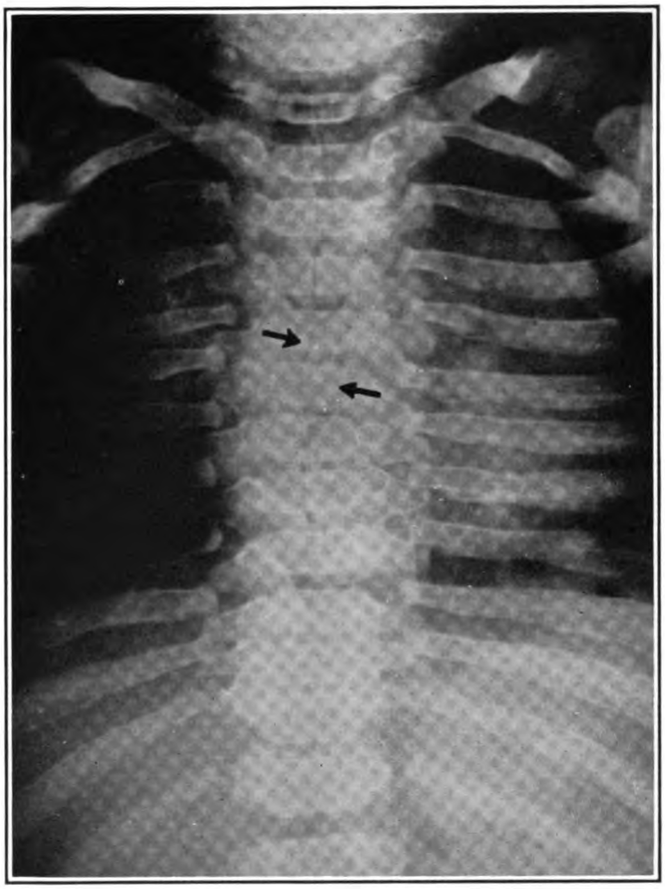

FIG. 79.—Unverified case of diastematomyelia, with malformed neural arches at the fifth and sixth thoracic vertebræ. A spur projects in the midline (arrows). The interpedicular spaces are widened in the midthoracic vertebræ, and the lower thoracic vertebræ are flattened.

Roentgenographic examination of the spine in well-advanced cases of diastematomyelia discloses fairly characteristic changes. These consist of a wide variety of malformations of the vertebral bodies, including hemivertebræ, unsegmented and hypoplastic vertebral bodies, narrow intervertebral spaces, kyphosis and scoliosis. The neural arches may be malformed, and lack of fusion and spina bifida is frequent. The interpedicular spaces in the involved areas may be markedly widened and the pedicles themselves may be flattened and thin, simulating the appearance which may be associated with a larger mass. The condition is often accurately ascertained by the demonstration of a spur which protrudes from below upwards between the split spinal cord or cauda equina as a bony septum which may measure up to 1.5 cc. in length. It may be seen as a rather spindle-shape or oval bony density in an area with malformation of the vertebral bodies and neural arches. This bony spur may be attached to the dorsal aspect of the underlying vertebral body with its apex directed into the lumen of the canal. While this process is usually single, it sometimes is multiple. The bony spur may be absent, and in its place a band of connective tissue which sometimes contains cartilage may protrude between the divided segments of the spinal cord. This connective tissue septum may be quite long, and within it a bony spicule may be present. According to Neuhauser, Wittenborg and Dehlinger the involved pedicles are not narrowed, differentiating the change from that due to tumor pressure. However, Walker (1944) observed that in 1 of his cases thinning of the pedicles was present, so that one cannot completely rely upon their configuration in differential diagnosis. Certainly the diagnosis of diastematomyelia would appear to come to mind quickest in the presence of advanced vertebral deformities, particularly if a bony spur in the mid-line can be demonstrated.

Pantopaque myelography often is helpful in diagnosing diastematomyelia, particularly if the characteristic spur is absent. In such cases the myelograms disclose a radiotranspartent midline linear shadow pointing towards the existence of a possibly unsuspected cartilaginous or fibrous septum. Associated with this may be irregularities in the pantopaque column secondary to arachnoidal adhesions. An unusual myelographic picture was mentioned by Cowie (1951) in his report of a twelve-year-old boy who had a spina bifida extending from the seventh cervical vertebra down to the mid-thoracic level together with a mild cervicodorsal scoliosis. The bodies of the second and third dorsal vertebræ presented irregular bony outlines mainly toward their left side with wedging of the lateral aspects of these vertebral bodies to a mild degree. A central defect in these vertebral bodies was noted together with considerable narrowing of the second and third intervertebral spaces without actual fusion. The interpedicular measurements in the upper six dorsal vertebræ were increased. Myelography showed a constant filling defect on the left side of the level of the second dorsal segment. This defect was irregular and confined to the lateral aspect of the column. At operation this change was found to be due to absence of filling around one part of the divided cord. The myelograms may also portray graphically the increase in the width of the spinal canal. An interesting observation was made by Walker (1944) who reported that a pneumomyelogram on 1 of his cases showed marked enlargement of the sacral sac.

Mention should be made of associated congenital anomalies which may be identified on physical examination. Cutaneous defects in the region of the underlying neural abnormality in the form of abnormal tufts of hair, dimples of the skin, lipomatous masses and port-wine angiomas should sharpen the observer's vision for the detection of the characteristic bony spur in the middle of a widened spinal canal.

The diagnosis is of more than academic importance inasmuch as surgical intervention sometimes affords considerable relief. Such cases are reported by Matson, Woods, Campbell and Ingraham (1950).

Diastematomyelia is often accompanied by evidence of neural deficiencies of the lower extremities characterized by loss of sphincteric control, failure to walk properly as the child grows, and abnormalities of the lower extremities including muscular asymmetry and malformations of the feet. Reflex changes may also be present in these regions. Walker (1944) asserted that the tension on the spinal cord produced by the fixation of the cord to the involved area is the main reason for symptoms developing early in adult life.

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Meningocele and Myelomeningocele.—By definition a meningocele is a protrusion of the meninges through vertebral or cranial defects occurring in infants as a congenital malformation. Depending upon the formation of the sac and its content a variety of names suggesting the histopathologic components have been suggested. That proposed by Recklinghausen in 1886 is considered as being the most practical. The condition is categorized as spina bifida with meningocele where the meningocele is associated with an incomplete fusion of the neural arch. The overlying skin may be entirely normal or covered by a thin parchment-like epithelium. The sac in these cases is composed of the meninges and contains only spinal fluid. Spina bifida with myelomeningocele indicates the presence of portions of the spinal cord or the nerve roots within the sac or attached to its walls. Many degrees of involvement of the concomitant neural tissue with the meningeal sac may exist, and a large number of confusing terms have been applied to this general condition. The term spina bifida occulta anterior with meningocele indicates that the protrusion may take place through a defect in a vertebral body or the sacrum with projection of the sac into the thoracic or abdominal cavities or the pelvis. In such cases the overlying skin may be normal in appearance except for local patches of hair or subcutaneous mounds of lipomatous tissue. When the sacs contain nervous tissue the condition is referred to as spina bifida occulta anterior with myelomeningocele.

The term "rachischisis" is generally accepted to imply a more advanced form of spina bifida with a large bony defect including absence of laminæ and pedicles, with direct exposure of nervous tissue and little or no sac formation. This condition is considered a developmental defect due to failure of fusion of the dorsal walls of the primitive ectodermal neural canal. Rachischisis may occur throughout or involve a portion of the spinal canal. The spinal cord and meninges in the areas of extreme involvement may be

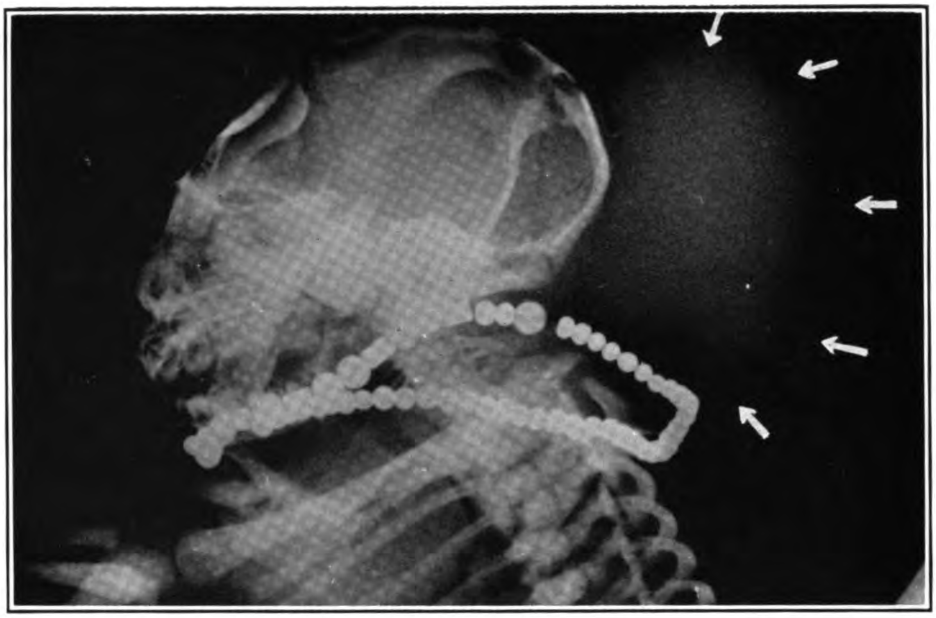

Fig. 80.—Lacunar skull with large myelomeningocele (arrows) projecting from defect in upper cervical spine.

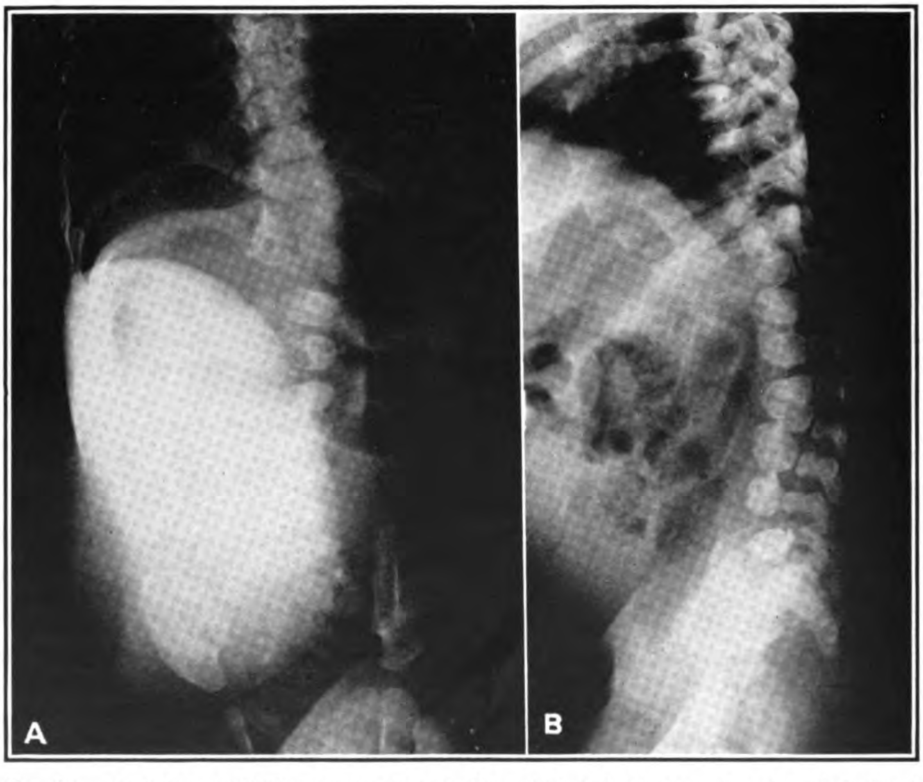

Fig. 81.—*A*, A large soft tissue mass projects from defects in the neural arches of the lumbar vertebræ. Numerous malformed vertebræ are present. Note the cleft, asymmetrically formed lower thoracic vertebræ and the anomalous ribs adjacent to the meningocele. *B*, Same case, lateral view. Note the malformed lumbar vertebral bodies. There is a dorsal convexity of the thoracolumbar spine.

greatly malformed or entirely absent. In partial rachischisis the bony defect usually is more pronounced in the neural arches and the degree of concomitant nervous tissue malformation is less pronounced.

A meningocele is not purely a simple prolapse of spinal membranes, but also is in the nature of a congenital malformation involving the soft tissues of the back including the skin and muscles, the bony structures of the spinal column and ribs and the spinal cord. The histologic components of the sac reflect ectodermal elements. The covering of the sac is skin which

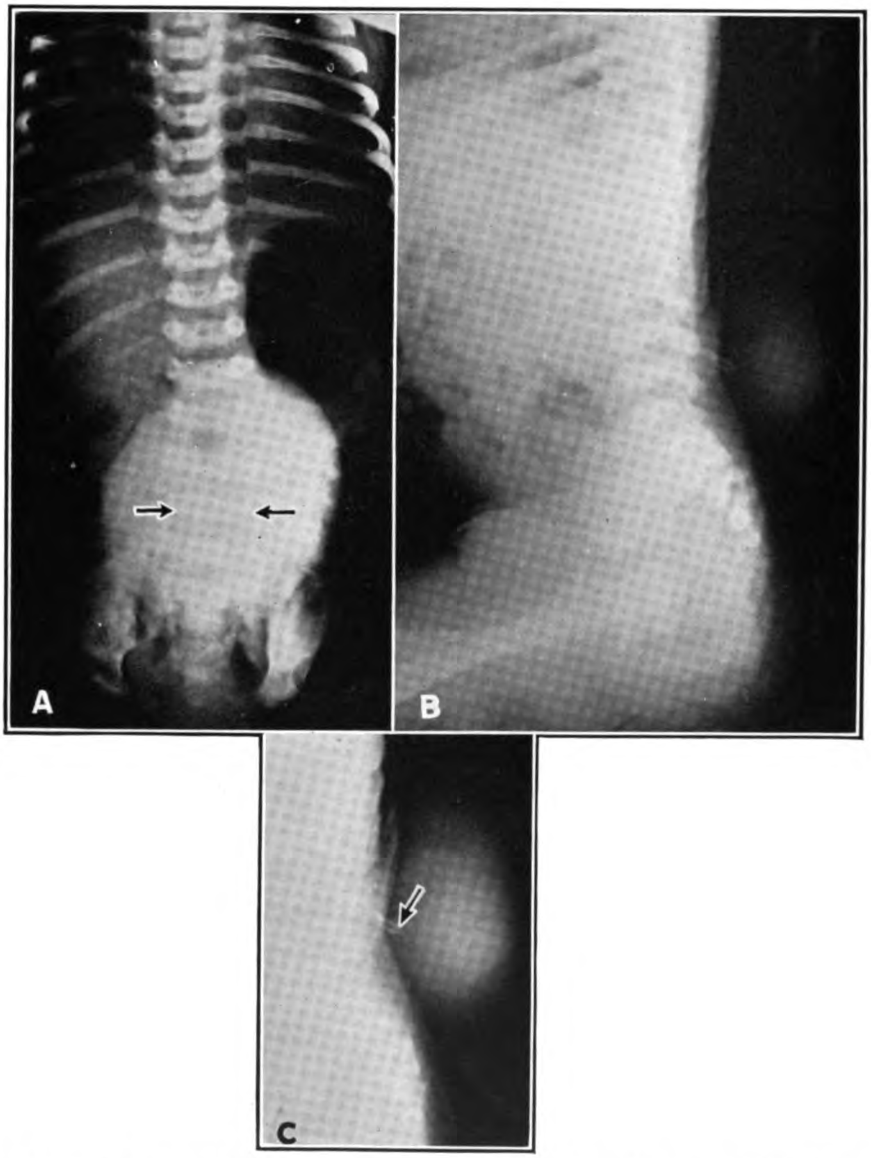

FIG. 82.—*A*, A large myelomeningocele overshadows the lumbar spine. The pedicles of the second to fifth lumbar vertebræ are spread (arrows). *B*, Same case, lateral view. The bodies of the lumbar vertebræ are well formed and no change in alignment is present. *C*, Same case. Soft tissue lateral spine roentgenogram showing meningocele. An anomalous bone spur, probably representing a rudimentary spinous process is present (arrow).

may vary from a normal appearing cutaneous structure to a thin, glistening silky integument. Usually the meningeal sac can be separated from the skin although this may be difficult when the skin is parchment-like. The sac usually is continuous with the dura mater and the dural and arachnoid coats become continuous at the site of the defect in the spine. Beyond this point the sac continues as a relatively undifferentiated meninx which may or may not contain nerve roots or spinal cord tissue. The defects in the spine which permit this abnormal development likewise permits a free exchange of cerebrospinal fluid between the cavity of the sac and the spinal canal. It has been pointed out that the association of hydrocephalus with meningocele indicated that the sac was part of an absorbing mechanism and that its total removal might be followed by progressive hydrocephalus.

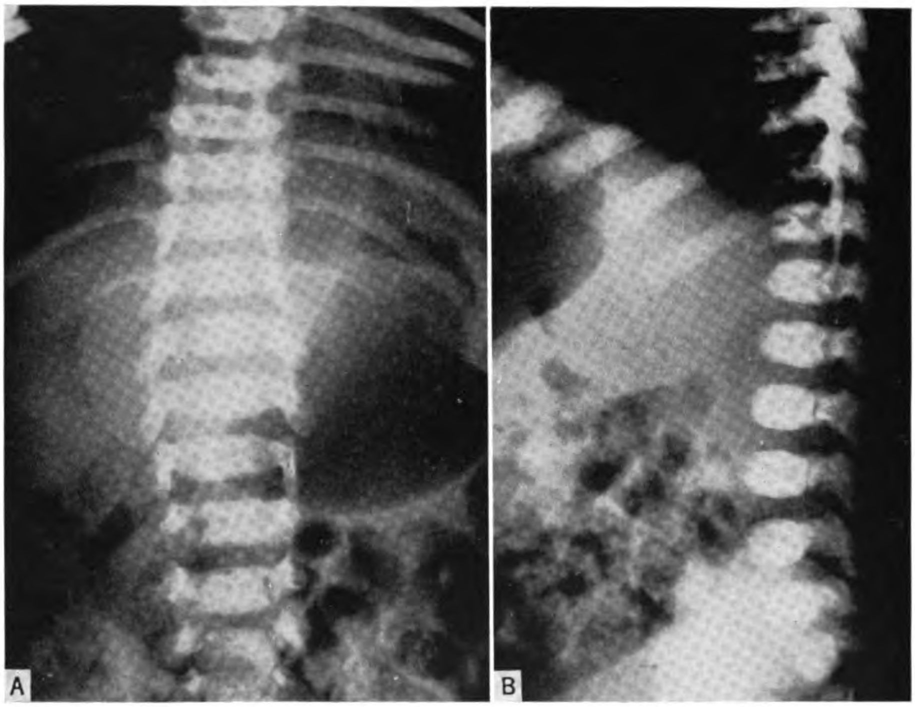

FIG. 83.—*A*, Myelomeningocele in a newborn infant. The meningeal sac is seen to the right of the lower thoracic and upper lumbar vertebræ. The pedicles are widely spread and the fusiform deformity extends from the tenth thoracic to the fifth lumbar vertebræ. *B*, Same patient, lateral view. The vertebral bodies are intact, and the intervertebral spaces are normal.

While in mild cases meningoceles may be unassociated with other skeletal anomalies, the concurrence of additional defects is quite frequent in moderate or advanced cases. Among these are included lacunar skull, hydrocephalus, the Arnold-Chiari syndrome, multiple congenital defects in the vertebral bodies, congenital dislocation of the hips, malformations of the viscera, including congenital anomalies of the kidneys, the gastrointestinal tract and of the lungs. Some of these have been considered under the section on vertebral malformations which in themselves are part of the picture of meningocele discussed here.

At this point it may be stated that the existence of a bifid spine is not absolutely necessary for the development of a meningocele. This is particularly true in the dorsal and sacral regions where the protrusion may occur through an intervertebral foramen or through an incompletely developed sacral segment, with absence of its anterior component and protrusion of the hernial sac into the pelvis.

Meningoceles occur most frequently in the lumbar and lumbosacral regions. The incidence of cervical and thoracic meningoceles is less frequent, and high cervical occipital meningoceles are even less common. In a series of 385 cases reported by Moore, (1905) 23 per cent were sacral, 34 per cent lumbar, 29 per cent lumbosacral, 4.5 per cent thoracic, 9.5 per cent cervical and 2 patients had occipital meningoceles.

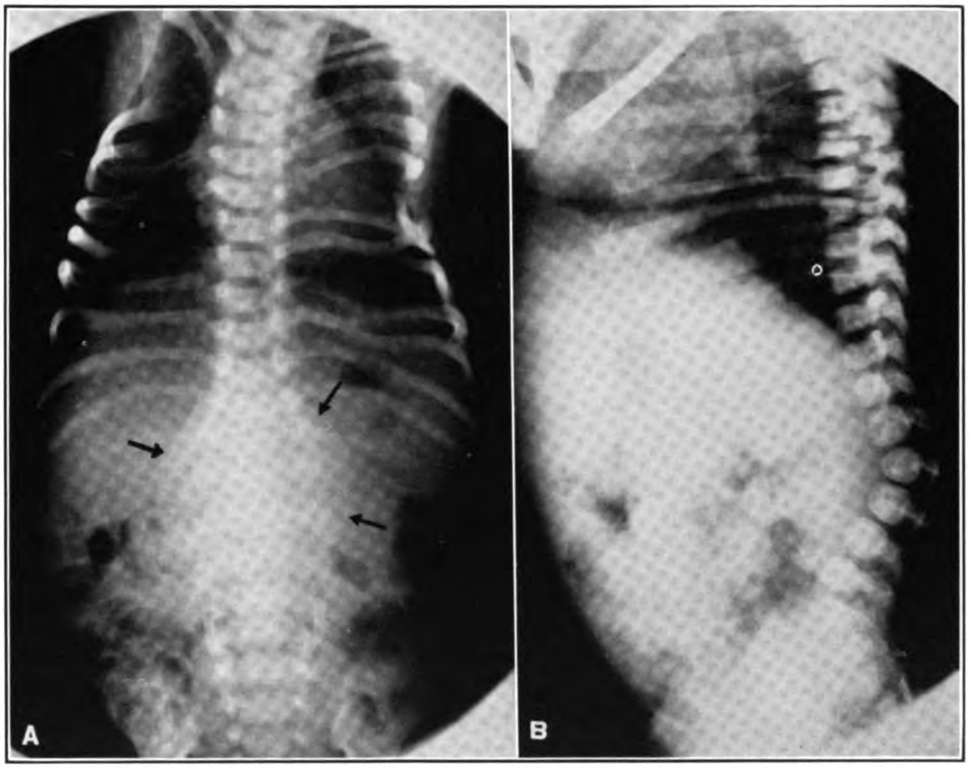

FIG. 84.—*A*, Myelomeningocele of thoracolumbar spine (arrows). There is a fusiform spreading of the pedicles of the lowermost two and all the lumbar vertebræ. Note the congenital rib malformations and the deformity of the upper thoracic vertebral bodies. *B*, Same case, lateral view. There is a local convexity of the upper lumbar spine. The body of the first lumbar vertebra is rounded anteriorly and is displaced dorsad.

In most instances the diagnosis of meningocele is a fairly obvious one based upon the identification of the sac. The underlying bony defects are best demonstrable radiologically. In most instances of uncomplicated meningocele, or myelomeningocele of the lumbar, sacral or lumbosacral regions one will encounter varying degrees of spreading of the pedicles beneath the area of the protruding sac. The area involved may be only one or two vertebræ, or more commonly the pedicles of several vertebral segments are involved. The spreading is usually arranged in a fusiform fashion, the widest portion of the interpedicular area being at the middle of the lesion. Immediately above and beneath the interpedicular spaces approach normal as the upper and lower extremes of the defect are reached. Associated with this is a maldevelopment of the neural arches of varying degrees, so that radiologically the laminæ and spinous processes are very difficult to identify. The mouth of the protruding meningocele does not necessarily correspond to the area of maximum widening of the interpedicular spaces. As a rule this region is considerably wider than the anatomic neck of the sac.

The appearance of the pedicles varies considerably. In some cases they may be relatively flattened but more often they retain a rather attenuated but still circular appearance. One gets the impression that the vertebral body seen through the sac is presented in much better detail than the supra- and infra-adjacent segments in which the neural arches are intact. In some cases the intervertebral discs spaces are relatively well maintained and the vertebral bodies are well formed. The concurrence of congenital malformations of the vertebral bodies and changes in alignment is by no means uncommon and may be accepted as part of the picture of meningocele or myelomeningocele.

On lateral views the soft tissue components and osseous contents of the protruding sac can be well demonstrated, and are helpful in recording configuration and size. The sac may also be seen on the anteroposterior views overlying the spine as a faintly radio-opaque shadow with a smooth outline. A radiologic diagnosis of meningocele *in utero* might be postulated if roentgenograms show a curvature of the lumbodorsal area convex dorsally, or if defects in the vertebral column are seen.

In the less frequently encountered meningoceles of the cervical and thoracic regions, a similar type of defect of the neural arches is noted, but as a rule the change is less marked. It is hard to determine even with roentgenograms whether a meningocele protrudes from a defect in the posteroinferior aspect of the occipital bone, or from an associated anomaly of the neural arches of the first and second cervical vertebræ. We have encountered instances in which defects were present in the neural arches of the upper cervical segments together with maldevelopment of the inferior aspect of the occipital bone. In other cases the meningocele protruded from a circular defect in the middle of the lower portion of the occipital bone. In both circumstances, the external appearance of the hernial sac was very much the same. The concurrence of lacunar skull with these meningoceles is frequent.

Intrathoracic meningoceles are uncommon lesions characterized by protrusion of a sac of dura through an enlarged intervertebral foramen into the thorax, displacing the posterior aspect of the parietal pleura anteriorly. In configuration and extent they are rather similar to hourglass tumors, and their association with neurofibroma is relatively frequent. It is presumed that intrathoracic meningoceles arise from a primary dural defect associated with hypoplasia of adjacent bone. Congenital mesodermal aberration affects both the dura and the vertebra, and because about two third of the reported cases of intrathoracic meningoceles have been associated with vertebral anomalies a common origin is postulated. Intrathoracic meningoceles without concomitant neurofibromatosis has been reported, but in these cases also vertebral defects have been noted.

Intrathoracic meningoceles occur as unilateral, and occasionally as bilateral tumors. The diagnosis is facilitated by the identification of associated spinal deformities such as kyphosis, scoliosis, vertebral erosions, enlarged intervertebral foramina, or scalloping of the adjacent vertebral bodies and ribs. Other malformations such as fused vertebræ or hemivertebræ with aplasia of the corresponding ribs also occur. An intrathoracic meningocele should be suspected when a mediastinal tumor is associated with neurofibromatosis or spinal anomalies.

It has been observed that in a few cases of intrathoracic meningocele the amount of cerebrospinal fluid which can be removed is unusually large. The diagnosis can be established with certainty by means of myelography. The use of pneumomyelography is particularly applicable, the demonstration of air entering the thoracic sac being of basic diagnostic significance. Similar observations also are possible with pantopaque. Unless clearly indicated by the patient's condition, surgery is considered inadvisable because of the danger of secondary infection.

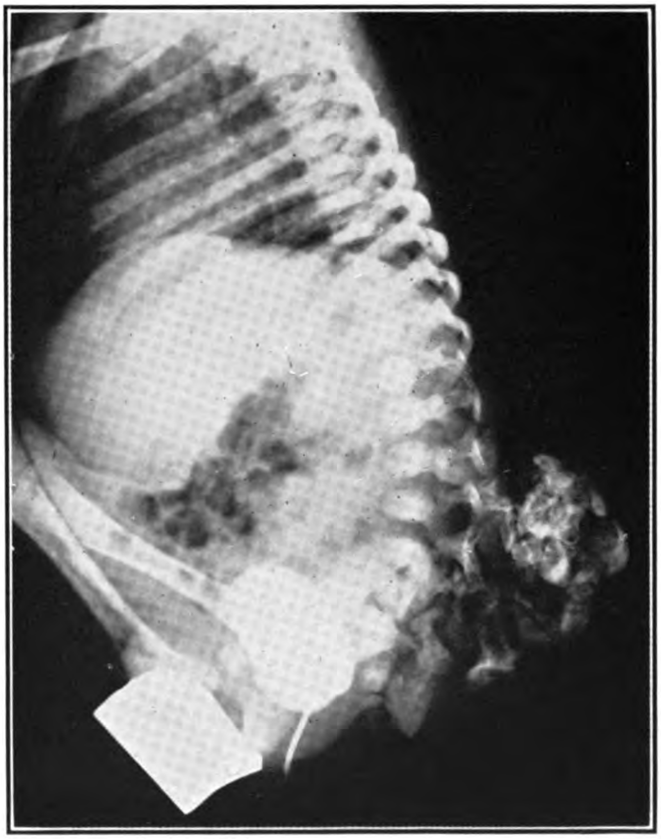

FIG. 85.—Lateral roentgenogram of case of payopagus parasiticums. The protrusion in the dorsal aspect of the lumbosacral spine was a meningocele containing a malformed twin fetus. (Courtesy of Drs. A. J. Segal and T. Gualtier, Coney Island Hospital, Brooklyn, N. Y.) Numerous other congenital anomalies, particularly of the genitourinary tract, were present.

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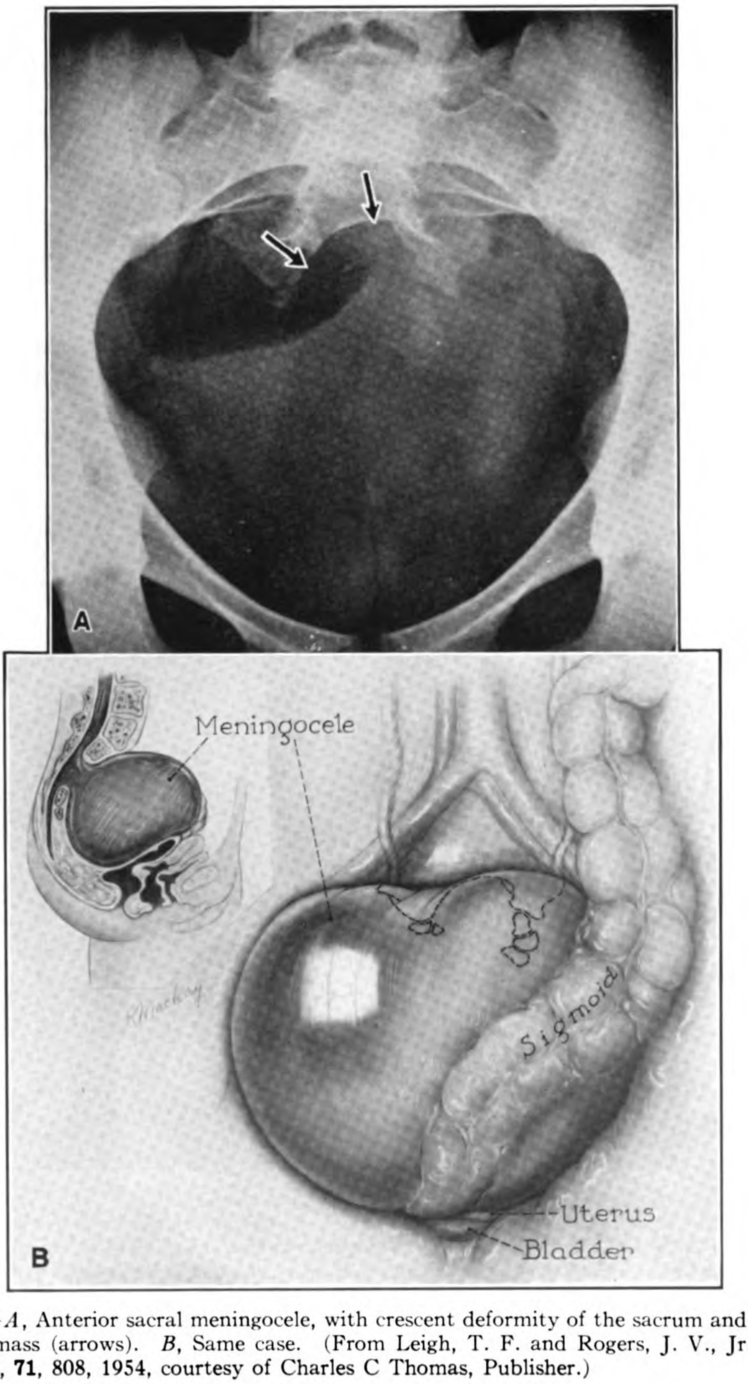

FIG. 86.—*A*, Anterior sacral meningocele, with crescent deformity of the sacrum and adjacent soft tissue mass (arrows). *B*, Same case. (From Leigh, T. F. and Rogers, J. V., Jr., Am. J. Roentgenol., **71**, 808, 1954, courtesy of Charles C Thomas, Publisher.)

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Anterior Sacral Meningocele.—This is a relatively infrequent congenital anomaly, usually associated with absence of half the sacrum producing a curvilinear, sometimes semicircular defect in either the right or left lateral halves. Projecting from this is a sac-like intrapelvic protrusion containing cerebrospinal fluid. Symptoms may be produced by pressure of the tumor on the pelvic viscera. Neurologic deficits may be incurred by involvement of the sacral segments, and a bizarre and most interesting symptom is the development of headache from intermittent increases in intracranial pressure by direct hydraulic action (Brown and Powell, 1945). Rarely agenesis of the lower sacrum may be bilateral rather than unilateral.

In addition to the plain films, which in themselves may be diagnostic, further information may be obtained by myelograms in which the pantopaque can be demonstrated entering the meningocele.

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Occult Meningocele.—This term refers to a widening of the distal end of the dural sac, usually unassociated with gross maldevelopment of the adjacent bone other than a variable widening of the sacral canal. The diagnosis is usually established myelographically. The caudal sac, instead of showing a pointed distal end, widens out into a spherical or cylindrical bag which may vary from 1 to about 5 cm. in diameter. The defect usually is enclosed within the vertebral canal, which sometimes may be demonstrable on lateral roentgenograms of the sacrum as a thinning of its dorsal aspect at approximately the second, third and fourth segments. The cyst usually is connected with the spinal subarachnoid space by a small channel. However, in several cases which we have observed the channel leading into the meningocele was quite wide, being a centimeter or more in diameter.

Occult or intrasacral meningocele often is asymptomatic, but has been associated with backaches and sacral nerve pain. The 3 cases reported by Archer, Cooper and Cimmino (1948) presented widely differing symptoms consisting of pain after trauma, urinary incontinence and various neurologic defects. Physical examination as a rule will not disclose a mass in this vicinity, but a congenital lipoma may be observed just beneath the skin or some of the deeper structures. Pool (1952) described traction effects on the spinal cord and local signs produced by occult sacral meningoceles in adults, and relief followed surgical intervention. At operation the sac was found to be lined with arachnoid, with nerve roots incorporated in its wall.

Communications with the spinal canal was by way of a narrow neck. One of his cases had an associated lipoma. The others had no lipoma, dermal sinus or aberrant cord. The filum terminale merged with the dura, forming the upper end of the meningocele. This was so taut that any traction resulted in a visible stretching effect.

This condition should be differentiated from the small outpouching sometimes observed on myelograms along the courses of the lower lumbar and first sacral nerve roots. These present themselves laterally as oval or sometimes circular shadows about a centimeter or so in diameter, usually located close to the axillary pouches and are considered to be arachnoidal diverticulæ. Rarely one of these may become large enough to simulate an occult meningocele.

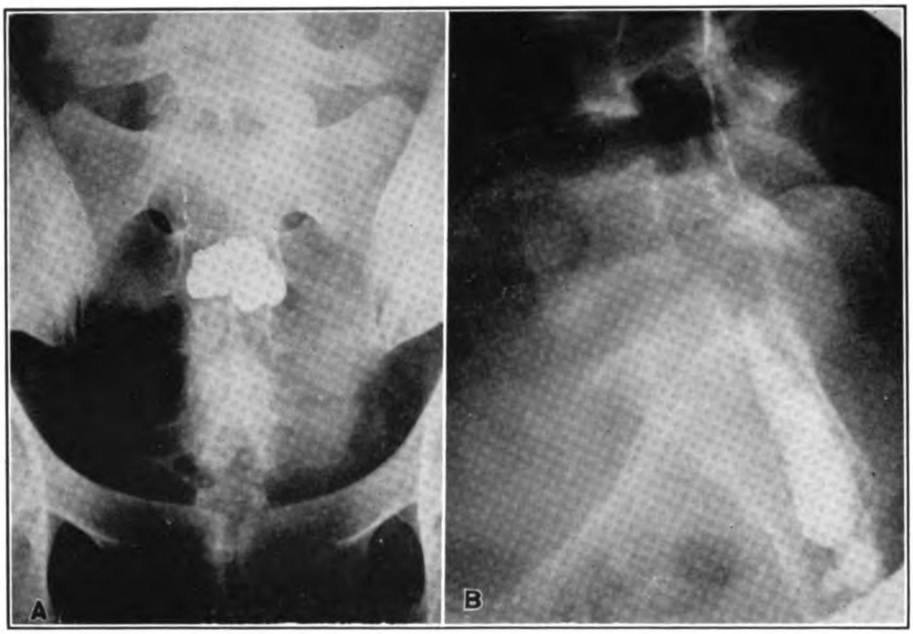

FIG. 87.—Occult sacral meningocele as seen on anteroposterior and lateral myelograms. The lateral film demonstrates the widening of the sacral canal.

SACRAL AND SACROCOCCYGEAL MALFORMATIONS.

Complete agenesis of the sacrum is an infrequent condition which may be recognized clinically by narrowing of the pelvis and shortening of the intergluteal fold because of absence of the spinal column below the lumbar region. Atrophy and dimpling of the buttocks are prominent, and the muscles of the lower extremities become atrophied. The muscular deficiency may become increasingly prominent as growth procedes. The condition develops early in fetal life, ossification of the sacrum beginning at about the eighth or ninth week. Concomitant defects are frequent. Included among these are dislocated hips, club feet, vertebral anomalies such as wedged or hemivertebræ, synostosis of the ribs and congenital subluxation of the knees. The caudal sac in these patients may be unusually high, sometimes making lumbar puncture difficult. The cauda equina also may be compressed, resulting in vesicorectal difficulties and variable motor and sensory disturbances. Radiologic diagnosis of this condition is readily established by noting the absence of the sacrum or a major portion of this bone, and close apposition of the iliac bones. With a more advanced form of this abnormality agenesis of the lower lumbar vertebræ has been observed as well.

Sacrococcygeal malformations also exist in a milder and considerably more frequent form. In these patients the coccyx and a variable number of lower sacral segments are absent. Not infrequently the only complaint

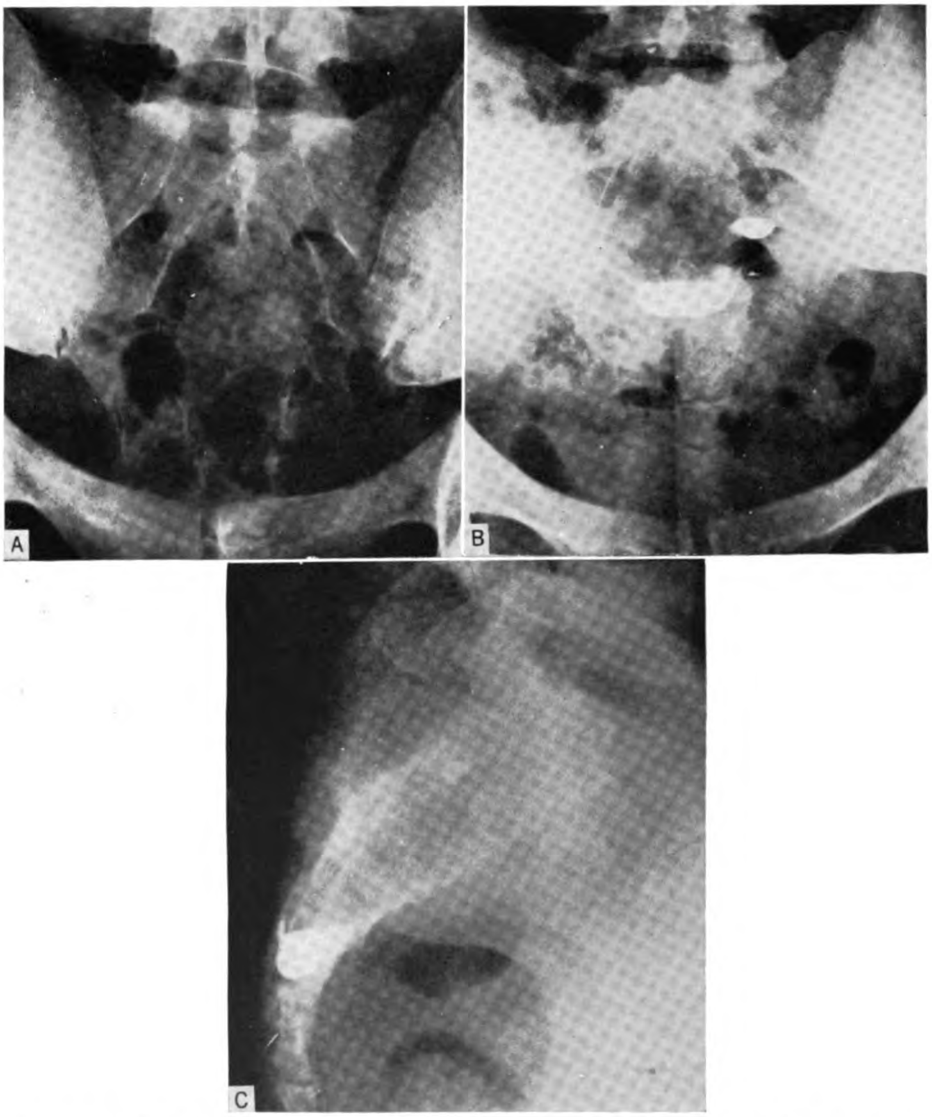

FIG. 88.—*A*, Tilt-up view of sacrum. The sacral foramina are spread, and a linear somewhat fusiform deformity is present in the sacral midportion. *B*, Same patient, erect myelogram. Pantopaque rests in the inferior aspect of the large sacral occult meningocele. *C*, Same patient, lateral erect myelogram. Note the increased sagittal diameter of the upper sacrum, corresponding to the meningocele. Pantopaque outlines the inferior aspect of the sac. (Courtesy of Dr. Robert Miner, Great Neck, N. Y.)

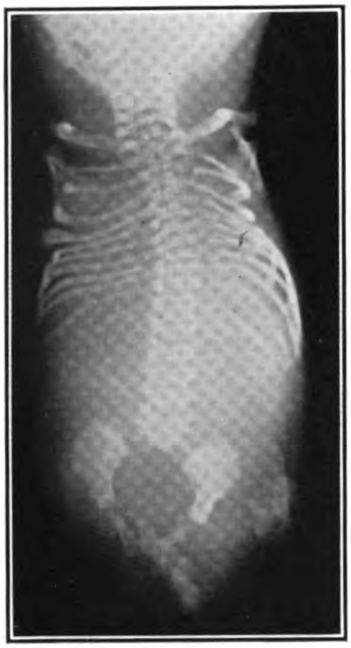

FIG. 89.—Absent sacrum in case of stillborn amelus. Additional deformities are seen in the ribs and thoracic vertebræ.

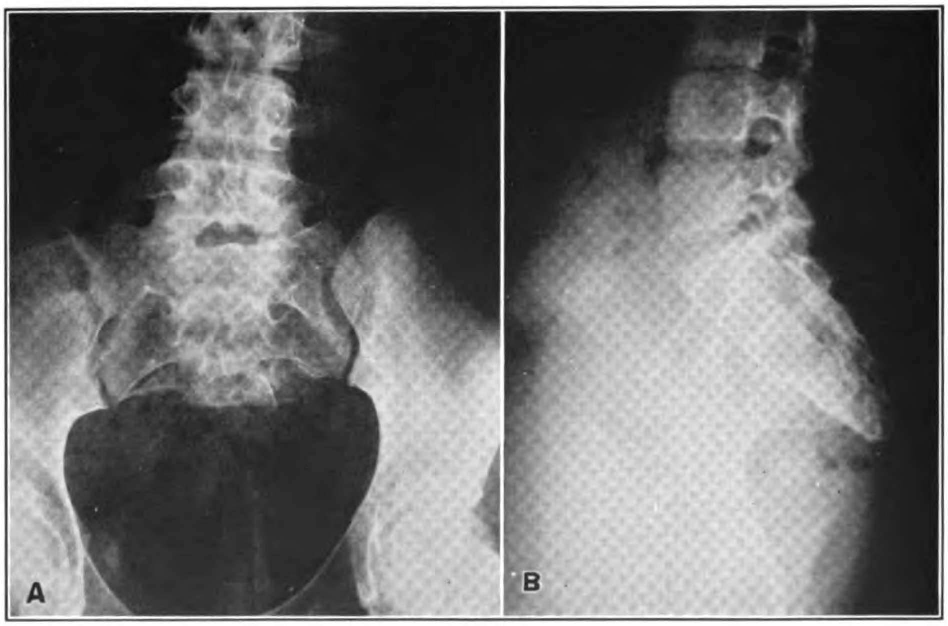

FIG. 90.—*A*, Congenital absence of lower half of sacrum. *B*, Same case, lateral view.

which brings the patient to the attention of the physician relates to mild urinary tract difficulties, sometimes with symptoms exaggerated because of superimposed infection. Rectal difficulties also exist, but usually are less pronounced. Neurological disturbances are mild or absent. However, concomitant anomalies of the vertebral column and of the viscera may occur. The identification of a sacrococcygeal malformation may serve to direct attention to the possibility of a concomitant urinary tract disturbance of anatomic or neurogenic origin.

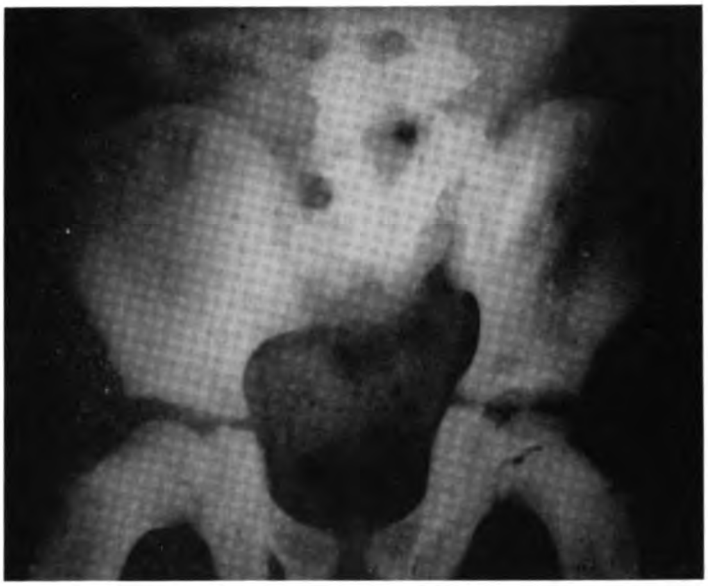

FIG. 91.—Congenital malformation of the sacrum resulting in approximation of sacroiliac joints. The left lateral aspect and the lower two thirds of the sacrum are deficient. Diagnosis made clinically because of narrowing of the hips.

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Pilonidal Sinuses.—Pilonidal sinuses are included among cases of cutaneous spinal dysraphism, inadequate or improper fusion of the embryonal tissues in the dorsal median region of the developing embryo, with resultant pathologic states manifested in the cutaneous, mesodermal or neural derivatives of these tissues. Dermal sinuses are regarded as malformations in the development of the neural canal.

On physical examination the diagnosis of pilonidal sinus is readily established by identification of the sinus opening in the midline between the buttocks overlying the lower sacrum or coccygeal segments. Associated with this opening may be a tuft of hair protruding from its orifice. The identification of these lesions point to further investigation of the bony structures underlying them. Sinus openings occur less frequently in the cervical or thoracic regions.

A connection between congenital dermal sinus and the dura was reported by Clifton and Rydell (1947), who presented what they considered the sixteenth such case. Their patient was twenty-two years old, which was older than usually seen. The plain films showed a spina bifida involving the entire sacrum. Myelography, however, showed that as the opaque medium flowed caudally it split into two columns. At operation this defect was found to be due to the thickened, taut filum terminale which was drawn and held posteriorly by inflammatory adhesions. This corresponds with the filum terminale syndrome described later by Garceau (1953) as occurring in many other congenital diseases such as spina bifida, meningocele and myelomeningocele, Klippel-Feil syndrome, cranium bifidum and diastematomyelia among others.

Pilonidal sinuses may involve segments above the lumbosacral area. A group of these was gathered by Kooistra (1942) who analyzed 14 cases from the literature including 1 of his own. One of these occurred in the cervical region, 5 in the thoracic region, 3 in the lumbar region, and 5 in the sacral region. In all but 2 of the cases described by him an associated spina bifida was present. Kooistra mentioned that in sinuses occurring over the upper half of the cord the picture of infection was usually absent; and that the clinical appearances were those highly suggestive of a spinal cord tumor. Cervical pilonidal sinuses may extend into the posterior cranial fossa, and when associated either with infection or an epidermoid, present the signs and symptoms of a cerebellar tumor.

In those cases which present symptoms referable to the spinal cord the advisability of myelography must be kept in mind. The discovery of a pilonidal sinus emphasizes the need for this procedure, which may either discloses deformity of the spinal canal, or partial or complete block.

The concurrence of epidermoid tumors and pilonidal sinuses will be discussed under the heading of Spinal Cord Tumors.

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Iliac Horns.—In 1946 Fong reported a condition which he designated as iliac horns. These were represented by symmetrical bilaterally placed protrusions from the posterior surfaces of the iliac bone adjacent to the sacroiliac joints. His case occurred in a twenty-seven-year-old woman and the processes measured approximately 2.5 cm. in diameter, and projected laterally and anteriorly for a distance of about 3 cm. He was unable to explain their etiology. This was later clarified by the work of Roeckerath (1951) who considered this deformity indicative of a hereditary dysplasia, associated in some of his cases with hypoplasia of the iliac bones, protrusion of the acetabulæ, coxa valga, dysplasias of the knee joints, elbows, feet, arms, hands and shoulder girdles and particularly of the nails. Anomalies of pigmentation of the iris also were seen. His work was confirmed by the reports of Cotette and Pfandler (1952) who reported on the radiologic and genetic aspects of this condition in a family comprising 51 subjects of four generations. They included spinal changes such as increased lumbar lordotic and cervical curvatures among the radiologic alterations.

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Congenital Torticollis —This condition was recently studied by Hulbert (1950), who reviewed 100 cases of torticollis and 117 cases of sternomastoid tumors. These were subdivided into postural and muscular groups. Congenital postural torticollis is present at birth and is not associated with sternomastoid tumor. The condition is transient and does not require operation for relief. Congenital muscular torticollis may be preceded by a sternomastoid tumor which is evident in about 20 per cent of cases on physical examination. Most of these respond spontaneously and leave no deformity. Excision in childhood is inadvisable. There are no definite associated skeletal changes in the cervical spine with either condition.

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Congenital Rotation of the Spinal Cord.—Morely (1953) reported a case in a paraplegic patient in whom the spinal cord at the level of the eighth and ninth thoracic vertebræ was found to be rotated about 60 degrees. The nerve roots were coiled half way around the cord above and below, with the dorsal root ganglion displaced into the canal. The left nerve root emerged from the right canal, and the right nerve root passed through the left canal. There were no roentgenographic changes associated with this condition.

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Spinal Curvatures.—The term scoliosis refers to lateral curvatures of the spine. Much depends on proper identification and management of these deformities. The clinical acumen and good judgement of the responsible physician often determines whether a patient undergoes one form of treatment or another, or indeed no treatment at all. Radiologic investigations must be accurate and the progress of the condition under continous observation to help in preventing progression to a distressing and disabling malformation in some instances.

The classification of scoliosis takes into account curvatures of known and unknown etiology. Accordingly, they are designated as due to congenital abnormalities such as osteochondrodystrophy, vertebral malformations such as, for example, hemivertebræ, or neurofibromatosis. Among other known causes are poliomyelitis, the various myopathies and neuropathies which result in muscular weaknesses and imbalances, and organic causes such as deformities which follow thoracoplasty. The larger group is of unknown origin, and is referred to as the idiopathic group. Some question has been raised as to a possible relationship between the myopathic and the idiopathic groups. A physiologic form also has been recognized.

The concept of primary and secondary curves is stressed by Cobb (1960) as basic to the understanding scoliosis problems. Following Ferguson's teaching, he points out that a primary curve is one produced by a deforming factor or force. A secondary curve is regarded as the result of involuntary muscular action to center the head on the pelvis as best as possible.

The normal spine as seen laterally presents four normal curves. The sacral and thoracic curves are concave anteriorly, and the cervical and lumbar curves are concave posteriorly. The sacrum, being a fixed structure incapable of movement, presents a structural curve which Cobb points out may be regarded as a primary curve. The thoracic vertebræ, being partly movable, has elements of both the primary (structural, fixed) curve and the secondary (functional) curves. The cervical and lumbar portions of the spine are capable of changing their configurations by bending, rotation or angulation. This permits the appearance of flexible, correctible curves which are regarded as functional (secondary) curves. Lateral curves are not normally present, but may form during motion, and then straighten out. In this sense, they are regarded as functional lateral curves. Some degree of this type of movement is present in the lower thoracic spine.

Normally the vertebral bodies are vertically aligned on a central axis, and are symmetrically formed. No changes in the configuration of the individual vertebræ are observed when the individual bends or moves. The alteration in the configuration of the spine is accomplished by the action of the intervertebral discs and the normal range of motion of the various vertebral segments. The presence of vertebral wedging and rotation is of primary importance in the identification of a structural curve. The angulation and position of the vertebræ likewise are extremely significant. Cobb is of the opinion that the terms "primary curve" and "secondary curve" might better be replaced by the designations "structural" and "functional" curve. Those with major structural changes can then be designated as major structural curves, those with minor structural changes as minor structural curves. Combined structural-functional curves are those which possess both structural and functional elements which the patient can correct when erect.

Physiologic scoliosis was investigated by Farkas (1941), who reported that measurements of the twelve thoracic and five lumbar vertebræ of 21 normal spines and 1 scoliotic spine in patients from six to eighty-six years old showed that the postnatal development of a normal spine became asymmetric at about the age of six years and progressed with advancing age. This asymmetry occurred in the vertebral bodies and pedicles as well as in the arches. The most striking feature was a dorsoventral elongation of the vertebral bodies. In the upper thoracic region this occurred predominantly on the left side, while in the lower thoracic region it was more prominent on the right side. In the lumbar region this change was again present on the left side. Lengthening of the pedicles on the side of the elongation and broadening of the pedicles on the opposite side was noted. Deviation of the neural arches and broadening of the pedicles opposite the side of the elongation were present. He felt that in physiologic scoliosis, contrary to the findings in pathologic scoliosis, wedging of the vertebræ was of only minor importance. He ascribed the cause of physiologic scoliosis to the human gait, which forces the spine into a threefold curve changing alternately at every step.

Idiopathic scoliosis accounts for 80 to 90 per cent of cases and may be familial in origin. Relatively few of these patients require surgical correction. The curvature of the spine is lateral. The primary curve is that which persists with the patient in forward flexion, and occurs anywheres in the spine. Compensatory curves are those above and below the primary curve, and usually straighten out in flexion. In lumbar scoliosis the primary curve is such that no compensatory curve forms below, so that only two are present. Cervical curves are uncommon.

Infantile idiopathic scoliosis makes up an important group. This condition begins somewhere before the second or third year, and is either resolving or progressive in its course. In the former group the curvature diminishes as the child grows, or the curve remains relatively unchanged. No treatment is required. With the progressive form of infantile idiopathic scoliosis the curvature usually is in the thoracic region with the convexity toward the left side. The condition supposedly is more frequent in males, and may progress rapidly to severe deformities or dwarfism. Idiopathic scoliosis also makes its appearance in juvenile or adolescent youngsters as a more rapidly progressive deformity. It has been stated that the lower the curve the better the outlook for the patient. Progression of the curvature is possible up until the time the veretbræ cease to grow. A relationship to the appearance of the iliac apophyses has been noted, vertebral growth ending when these appear. After this time the curves do not progress much. Curvatures of the thoracic spine also occur toward the right side, but supposedly are later in onset. The factors which appear to influence the prognosis include the length of the curve, the degree of vertebral rotation at the apex, the age of onset and the rapidity of deterioration.

In idiopathic scoliosis a more complex deformity exists, consisting of lateral deviation of the spine with rotation of the vertebræ. As a rule, the curves present with idiopathic scoliosis are multiple. A single major curve accompanied by one or two compensatory curves is observed fairly frequently, the curves compensating for the spinal vertical alignment. Single major curves are more likely to cause structural deformities. Idiopathic scoliosis may involve the lumbar spine alone beginning at about ten years of age and continuing to adolescence. This type, as a rule, is innocuous, but may be followed by degenerative hypertrophic changes of a painful nature later in life. Involvement of the thoraco-lumbar region alone is infrequent. This curvature usually develops during adolescence, and may be deforming. It requires careful observation. Scoliosis limited to the dorsal spine may be severe and rapidly progressive, requiring expert evaluation and management. It occurs mainly in girls from five to eight years of age.

Opinion is still divided as to whether the lateral deviation or the rotation of the vertebræ is the initial deformity, whether these two changes are independent of each other, or whether they are the natural concomitants of some underlying cause. According to Thomas (1947) it is generally assumed that rotation is secondary to lateral deviation. The latter element is always accompanied by some rotation except in the mildest grades, while with more severe curvatures the degree of rotation is more pronounced. No constant ratio exists between these two components. The rotational defect appears to be the more resistant to correction.

The structural changes in the vertebræ responsible for this change in alignment is principally the development of a wedge-like deformity of the vertebral body. Accompanying this, and much more difficult to explain are the rotational defects of the involved vertebræ which appears as the lateral deviation develops. The rotation occurs on a vertical axis situated somewhere behind its spinous process. Rotary displacement may be recognized roentgenologically by the lateral displacement of the spinous process and by the general asymmetry of the shadows of the pedicles in relation to the corresponding vertebral bodies. The pedicles on the convex side of the deformity are placed medially away from the lateral borders of the bodies on that side, while the pedicles on the concave side become less distinct and in some advanced cases may disappear from view entirely. The neural arches of the rotated vertebræ likewise become deformed, and the articular facets on the concave side are thrown out of alignment with those on the convex side, with consequent locking of the vertebræ in their rotated position.

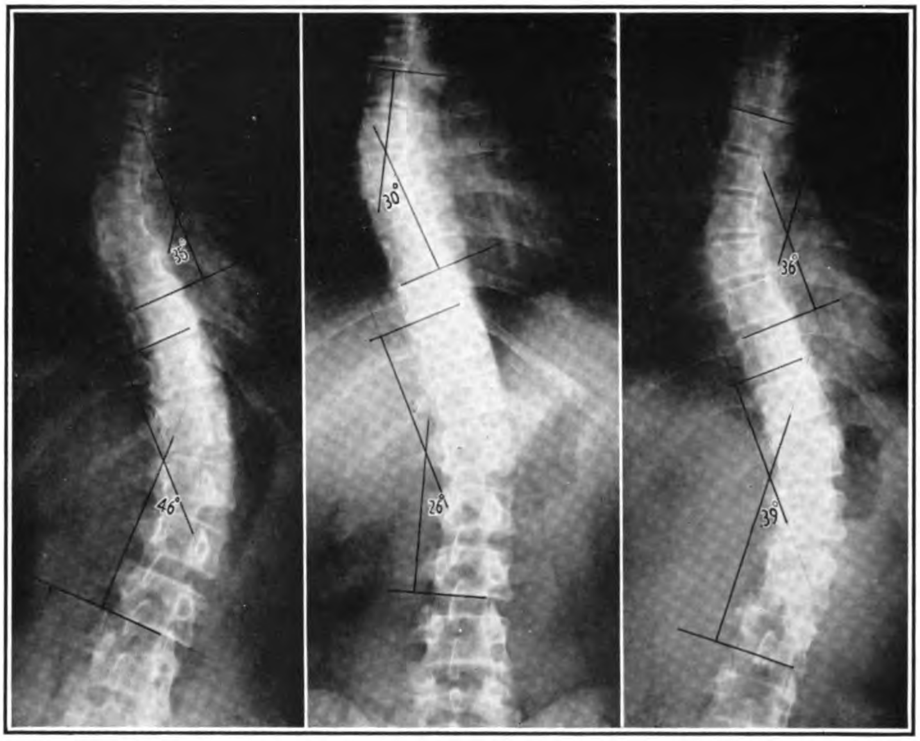

FIG. 92.—*A*, Scoliosis study, erect posture. *B*, Same case, supine position *C*, Same case as *A* and *B*, sitting position.

The most deforming curves originate early in life, and it has been noted that scoliosis appearing later in adolescence usually has a better prognosis.

The curvature usually ceases to progress about one year before ossification is complete. According to Ponseti and Friedman (1950) a rapid increase in the curve as noted on serial roentgenograms is of poor prognostic significance. James (1951) stated that only 5 to 10 per cent of idiopathic curves become severe enough to require correction and fusion.

According to Arkin (1949) wedging can best be explained by unilateral epiphyseal arrest. This had also been brought out experimentally by Bisgard and Musselman (1940), who produced scoliosis by destroying only one side of a growing vertebra, so that wedge-shaped vertebral bodies and lateral deviations of the spine were produced. Experimental scoliosis was also studied by Nachlas and Borden (1950) who produced a human-like type of scoliosis in dogs by experiments which controlled epiphyseal growth with piano wire staples. The deformities were progressive even after the staples had been removed. Compensatory thoracic spine curves appeared in several animals. The relationship of the growing epiphysis to scoliosis was brought out by Haas (1939) who in dog experiments was able to produce kyphosis and scoliosis by injuring the cartilaginous vertebral plates. He concluded that the growth in the length of vertebræ occurs in a manner similar to growth in length of long bones, namely, by a proliferation at the cartilage end plate with no interstitial growth within the body. Somerville (1952) reported that so-called structural scoliosis was the result of failures of growth of the posterior elements of a segment of the spine. He presented the results of experiments in young rabbits in which trauma to the laminæ and spinous processes was followed by curvature of the spine.

Radiologic investigations of the spine in cases of scoliosis should include films which visualize the spine from the level of the iliac crests up to the cervical region. These studies should be made with the patient in the supine, the sitting, and in the erect positions. In the event of a significant degree of curvature, further studies may be made with the patient seated with a 2- to 3-inch block under each buttock. Additional studies may be required with the patient erect, first with the right and then with the left foot elevated by a 2- to 3-inch block as deemed necessary. The study should be augmented by lateral films as well as conedown studies of the lumbosacral articulation in the anteroposterior and lateral views. Repeated examinations for changes in the scoliosis angles should be made at from three- to six-month intervals. The scoliosis angles may be identified as shown in Figure 92, p. 156. A chart may then be prepared in which the degree of wedging, rotation, angulation and the position of the vertebræ is recorded according to date, utilizing the same points of reference for each study as developed by Cobb (1948) whose "Outline for the Study of Scoliosis" should be read carefully in the original.

Paralytic scoliosis following poliomyelitis may appear within a few months after the acute episode, and be rapidly progressive. It may also be slow in development, appearing several years after the original disease. According to Cobb this group makes up 30 to 50 per cent of all cases requiring fusion, and is one of the more serious complications of the disease, requiring prolonged and expert management.

The weakening of spinal muscles by poliomyelitis results in structural and postural imbalances causing a wide variety of spinal curvatures. The superficial longitudinal muscle group controls extension, while the deep oblique and transverse groups affect rotary movement. Both groups participate in lateral flexion. The extraspinal muscles also participate in the establishment of normal posture, and departures from their normal action because of poliomyelitis as well as changes in the alignment of the pelvis contribute to the development of spinal curvatures. Poliomyelitis first causes paralytic effects, followed by scoliosis. Functionally the paralytic spine has increased mobility with a high degree of flexibility and compressibility, with resultant loss of the thoracic kyphosis and lumbar lordosis and translatory shift of the vertebræ. With relatively mild disturbances, a generalized relatively slight thoracolumbar "C" curve develops which improves on recumbency. More advanced changes produce practically a collapse of muscular tone, with extensive, mobile thoracolumbar curvatures which include rotational deformities as well. As a rule a primary curve appears first, which is fixed and usually the most pronounced. Sometimes a double curve appears.

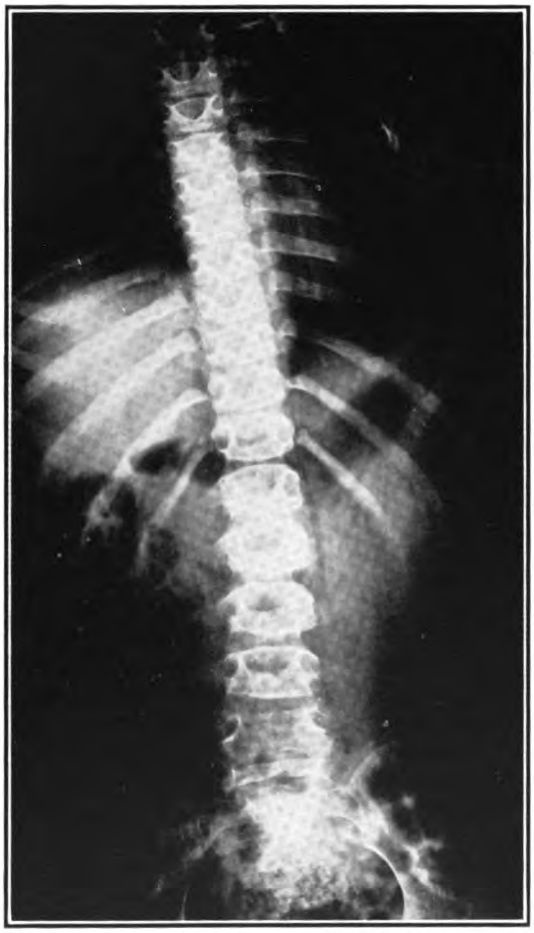

FIG. 93.—Anteroposterior supine view of thoracic and lumbar spine, post-polio curvature.

Radiologically the involved vertebræ reflect the positional and nutritional changes incident to lack of movement and muscle tone. The bodies assume a somewhat clouded and demineralized appearance. The discs diminish in size, and are distorted in conformity with the changed vertebral bodies. Minute calcific flecks or somewhat large calcareous deposits may appear in the discs. The time required for these changes is variable, and years may go by before the final effects are attained. During this course the curvatures may change considerably.

Unusual forms of scoliosis have been reported because of weakness of the spinal muscles occurring in conditions such as pseudohypertrophic muscular dystrophy, arachnodactyly, progressive muscular dystrophies and dystonia musculorum deformans.

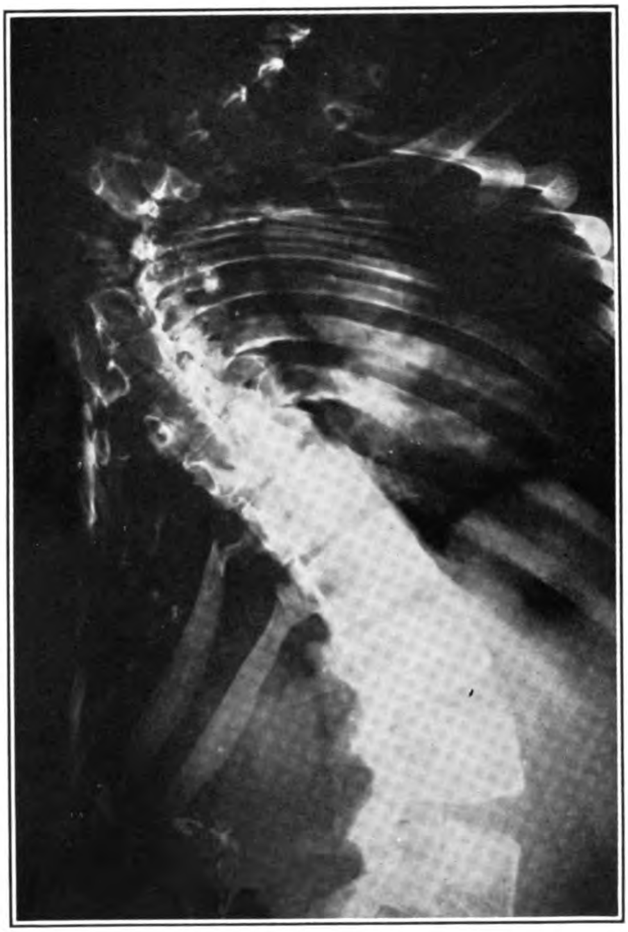

FIG. 94.—Post-thoracoplasty scoliosis convex to the right. Film made five years after operation.

Scoliosis may also follow empyema, the concavity of the spinal curvature being directed towards the site of the empyema, with relatively little or no rotation of the vertebral bodies, (Selig and Arnheim, 1939). Such changes may also follow thoracoplasty in which case the convexity occurs towards the resected ribs. The vertebræ in these instances develop little or no structural change, and the deformity may be corrected surgically by removing a portion of the thoracic wall which may release the pull of a pleural scar. Bisgard (1934) pointed out that such realignment of the vertebræ would be impossible in the presence of significant structural changes in the vertebral bodies.

Intensive radiation therapy directed over the spine also can produce scoliosis by suppressing vertebral epiphyseal growth. Arkin and Simon (1950) were able to produce structural scoliosis in rabbits by irradiating the vertebræ asymmetrically, resulting in wedging of the bodies with the lesser height towards the irradiated side. Arkin, Pack, Ransohoff and Simon (1950) reported a case in a thirteen-year-old schoolgirl who had been treated during infancy for pigmented diffuse nevi on which melanotic tumors had developed, which were later excised. When nineteen months old the patient was treated for a Wilms' tumor by means of preoperative *x*-ray therapy, nephrectomy and postoperative irradiation. Scoliosis appeared at the age of nine years and by the age of thirteen the curves were seen to follow exactly the fields of irradiation with the wedging directed towards the irradiated sides. They noted that in addition to the insufficiency of growth in the vertebral bodies, similar changes occurred in the twelfth left rib, a left lumbar transverse process and the left ilium, all of which had been included in the field of treatment.

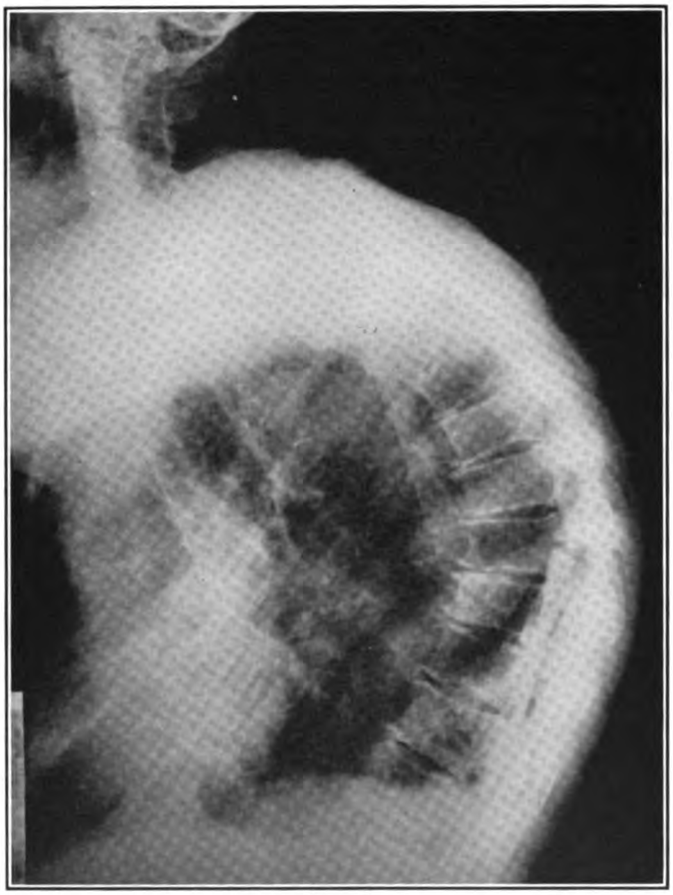

FIG. 95.—Lateral roentgenogram of dorsal spine in case of senile kyphosis. Note the relatively well maintained intervertebral discs and the slight spondylosis. The intervertebral discs are more narrowed anteriorly than posteriorly.

The effects of irradiation on the growing spine were summarized by Neuhauser, Wittenborg, Berman and Cohen in 1952, who reported on 45 patients who had received *x*-ray therapy over the spine. Thirty-four of these patients were living and had been observed over a period averaging six and one-half years. Postmortem examinations were available on 11 patients. The significant changes in these were nonspecific growth retardation and irregularity of ossification of the epiphyseal cartilages. These authors noted that scoliosis is only rarely produced if the spine received uniform irradiation.

A rare form of scoliosis noted in association with marked thymic enlargement was reported by Paltrinieri (1952) with recession of the curvature after *x*-ray therapy. He believed that the deformity occurred only with pressure on one side and that the curvature acted as an accommodation curve. Two cases of scoliosis associated with paravertebral teratoid tumor were reported by Lawson (1953).

Scoliosis Incident to Spinal Cord Disease.—It has been stressed by many that curvatures of the spinal column may appear during the course of the growth of a spinal cord tumor. In a review of 2000 cases of scoliosis of all types Ruhlin and Albert (1941) found 7 cases with symptoms referable to the central nervous system. Three of these patients had rachitic scoliosis, 3 congenital scoliosis and 1 was of doubtful origin, associated with neurofibromatosis. In no instance was cord involvement observed in paralytic or habitual types of scoliosis. The paralytic signs developed regardless of the direction of the primary curve, but all of the curves involved were in the cervicothoracic region and were sharply angulated.

In comparison with these changes in which the nervous system damage was due to the scoliosis, one may cite the cases reported by Boldrey, Adams and Brown (1949) in which scoliosis appeared as a manifestation of disease of the cervicothoracic portion of the spinal cord. They reported 10 such cases in which scoliosis was the first or an early principal sign. Included in this group were cases of cystic lesions of the cord, angioma, spinal varix, adhesions and extramedullary tumor. They mentioned that of a total of 60 consecutive surgically proven spinal cord lesions moderate to pronounced scoliosis was demonstrable in 15, with the convexity usually towards the side of the lesion where this was demonstrable. Williams and Stevens (1953) were of the opinion that any child with idiopathic scoliosis should have careful neurologic examination, and cited the case of an eight-year-old boy with gradual curvature of the spine, frequent stumbling and back pain, enuresis and progressive constipation in whom the plain roentgenograms were normal and myelograms revealed a block at the level of the tenth dorsal vertebra. Operation in this case disclosed an ependymoma.

Paraplegia complicating scoliosis has been noted in occasional cases and has been considered as due to the unequal growth of the dura and the spinal column in a tight dural canal. The exciting cause for the paralysis is the inability of a tight dura to accommodate itself to such growth (Kleinberg, 1951). In a review of 41 cases McKenzie and Dewar (1949) noted that the paralysis appeared during the period of rapid growth at a time when the scoliosis, if not controlled by treatment, is suddenly and markedly increased. At operation in their cases a linear thickening of the dura was noted which extended across the neural canal obliquely, compressing the cord. One patient had a spur of bone compressing the cord. The treatment in such cases is surgical. Myelography is of prime importance in establishing the site of compression, which is usually close to the apex of the curve. Most of the cases of this nature observed by the authors were idiopathic scoliosis, but others have been associated with rickets, poliomyelitis and neurofibromatosis of bone.

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Kyphoscoliosis and Kyphosis.—This condition is characterized by angular or curvilinear deformity of the spine in which convexity is directed dorsally or dorsolaterally, and occurs most frequently in the thoracic region. A large variety of pathologic states may be responsible for this malformation. It is most commonly due to faulty posture, to generalized weakness, poliomyelitis, congenital deformities, neurofibromatosis and occupational diseases. Various forms of osteoporosis, arthritis, rickets, Paget's disease, acromegaly, hyperparathyroidism, may be causative factors. All of these produce a more or less smooth curve which vary in degree from a mild dorsal convexity to an almost semicircular configuration. These conditions will be discussed under their respective causes. An angular kyphosis is usually incident to tuberculosis, syphilis, malignant disease, congenital anomalies of vertebral column or compression fractures. These too will be discussed under their respective headings. Rachitic kyphoscoliosis with spinal cord compression was reported by Ponsold (1935). Angular kyphosis of nontuberculous origin is relatively rare and according to Gulledge and Brav (1950) only 41 such cases have been described in the literature.

Kyphosis and kyphoscoliosis both may be associated with paraplegia due to compression of the spinal cord because of the angular deformity of the respective vertebræ or because of protrusion of bony spurs or fibrous bands into the spinal canal (McKenzie and Dewar, 1949). These lesions may be identified by means of myelography, in which case the demonstration of block is of great diagnostic importance (Kleinberg and Kaplan, 1952).

The development of other complications such as cor pulmonale (Samuelson, 1952), atelectasis, emphysema and bronchitis (Hertzog and Manz, 1943), and compression of the pulmonary vessels and mediastinum have been mentioned (Chapman, Dill and Graybiel, 1939), and (Kerwin, 1942). The presence of esophageal ulcers with kyphoscoliosis was reported by Haubrich (1953).

Lordosis.—In this condition there is an exaggerated convexity of the lumbar spine directed anteriorly. This may be produced by weakness of the back muscles from any cause such as poliomyelitis or muscular dystrophy, or it may result as a compensation for a kyphotic curve. Lordosis may also be associated with enlargement of the abdomen such as may occur with obesity, pregnancy, ascites or hepatosplenomegaly. An exaggerated form of lumbar lordosis may sometimes be observed in achondroplastic dwarfs and cretins. Probably the most frequent cause of lordosis is simply poor posture.

The radiologic changes associated with these various conditions are discussed under their specific headings. In those cases of lordosis in which there is only the element of poor posture or relaxation of muscular support, no gross changes in the vertebral bodies or their interspaces can be seen.

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Chapter

3

Diseases of Congenital, Endocrine or Metabolic Origin

CONGENITAL DISORDERS

Chondro-osteo-dystrophy (Morquio-Brailsford's Disease)

The nomenclature for osteochondrodystrophy is confused. Among the names which have been employed are familial osteochondrodystrophy, dyschondroplasia foetalis, chondrodysplasia, osseous dystrophy, atypical achondroplasia, rachitic chondrodystrophy, dysostosis multiplex, osteodystrophia fibrosa, chondrodystrophic rheostosis, congenital perversion of growth, and ecchondrodysplasia (Jacobsen, 1939).

Morquio-Brailsford's disease is a familial disease of unknown etiology resulting from a developmental defect in germ plasm, characterized by a hyperplasia of cartilaginous growth. Consanguinity may be a factor. In its most characteristic form osteochondrodystrophy is manifested by dwarfism, deformities of the bones of the trunk and extremities, including coxa vara deformities of the hips and genu valgum and pes planus deformities. The skull in the majority of cases remains unchanged, but some instances, in which enlargement of the cranium has been noted, have appeared in the recent literature (Feldman and Davenport, 1951). As a rule the intelligence of the afflicted person is normal and may even be above average. The association of mental defects with Morquio's disease was mentioned by Farrell, Maloney and Yakovlev (1942). The first classic descriptions of the disease were those of Brailsford (1928) and Morquio (1929). However, in 1920 Wheeldon had reported a case which he believed to be one of achondoplasia in which a wedge-shaped vertebra was mentioned as a prominent change.

The radiologic diagnosis of osteochondrodystrophy rarely may be made on roentgenograms taken during the third trimester of pregnancy. Brailsford mentioned that he had observed chondro-osteodystrophy of such a severe degree that the fetus died *in utero*. All of the bones of the skeleton with the exception of the clavicles in this case showed evidence of defective development. Caffey (1950) stated that neither the clinical nor the roentgen picture of Morquio's disease had been observed in fetuses or in infants. The disorder first becomes evident during the fourth and fifth years of life when difficulties in walking and deformities of the spine are noted by parents. In this connection it is interesting to note that the original case described by Wheeldon was first seen at the age of twenty-two months, and later was reported again by Pohl (1939) at the age of twenty-two years.

The usual picture of Morquio's disease is an infant who apparently is perfectly normal at birth, and develops fairly well until the age of from three to five years. At this time growth may cease, and dwarfism with malformations of the trunk and extremities becomes apparent, associated

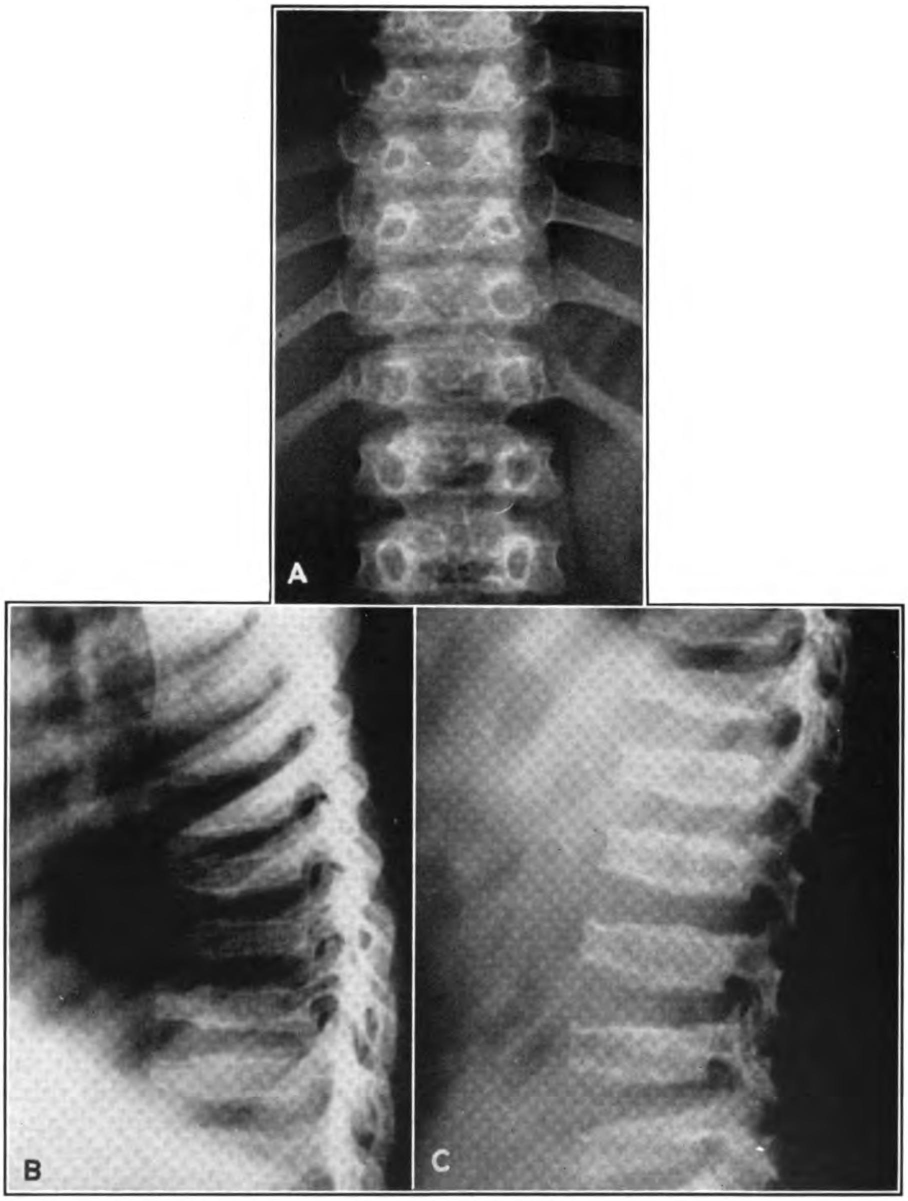

FIG. 96.—*A*, Osteochondrodystrophy in a fifteen-year-old female. Anteroposterior roentgenograms of thoracolumbar spine. The vertebral bodies are elongated and flattened. *B*, Same case. Lateral view of thoracic spine. The vertebral bodies are flattened, and a squared-off tongue-like protrusion is present at the anterior aspects of the lower thoracic vertebræ. The intervertebral spaces are widened. Note the increased discal heights at their anterior aspects. *C*, Same case. Lateral roentgenogram of the lumbar spine. The vertebral bodies are flattened. The tongue-like protrusions are less pronounced than in the lower thoracic vertebræ. The intervertebral discs are widened, particularly anteriorly. (Courtesy of Dr. G. Seaman, Brooklyn, N. Y.)

with progressive weakness. The full-blown appearance is that of a markedly shortened and deformed individual usually of normal mentality, with a short neck and trunk, kyphosis and deformities of the upper and lower extremities with some shortening. The chest is deepened and flattened with a very prominent sternum. A considerable degree of muscular weakness is present, which gives rise to alterations in gait and stance. The joints are large and have a peculiar hypermotility and laxity, so that the patient stands with hips and knees flexed in a crouching position, with the pelvis tilted forward and head sunken between the shoulders.

The variability in the changes associated with osteochondrodystrophy was stressed by Brailsford, who divided such cases into four groups. The first was that in which the disease was evident *in utero* or shortly after birth. The second group was that in which the active phase of the condition appeared to cease before puberty, giving rise to a moderate generalized form with marked shortening of the limb bones and pressure deformities of the extremities and the long bones and vertebral bodies. His third group was one with changes confined chiefly to the spine and hip joints, and less frequently to the knees. In the fourth group changes were confined to the vertebral column, with no irregularity or growth disturbances visible in other parts of the skeleton.

Osteochondrodystrophy is considered a probably hereditary developmental defect in the germ plasm, characterized by hyperplasia of cartilage growth accompanied by osteomalacic deformities of the skeleton. Radiologically this is manifested by shortening and distortion of the various bones of the extremities with non-union and distortion of the epiphyses and delayed ossification. The joint surfaces become widened and the articular surfaces are irregular, flattened and eroded. The acetabular and glenoid fossæ are shallow and irregular. The vertebral bodies rarely are involved in the sense that cartilaginous rests occur in their bony substance. However, Russo (1943) reported a case of a twenty-six-year-old dwarf seen in the seventh month of pregnancy in whom radiologic examination of the vertebral bodies disclosed numerous areas of translucency which he considered to be due to islands of cartilage which had failed to calcify.

The radiologic changes evident in the spine are among the important ones associated with osteochondrodystrophy. In its most developed form there is practically complete involvement of the vertebral column, the characteristic change being a pronounced flattening and widening of the respective vertebral bodies. In their anterior aspects the vertebræ present a peculiar tongue-like elongation of the central portion of the body. The superior and inferior articular surfaces of the vertebral bodies may be quite roughened, and anterior wedging of varying degrees is frequently observed. Multiple centers of ossification appear at the apophyseal rings, which later unite. These deformities of the vertebral bodies result in varying degrees of kyphosis, some of which are of an advanced nature. Occasionally the deformity is so great that a marked kyphosis with gibbus formation results. It may appear as if the vertebral bodies at the thoracolumbar junction had been squeezed out of line over the small vertebral body at the apex of the curve. These kyphotic disturbances are frequently accompanied by lateral curvatures of the thoracic and lumbar vertebral column. In the cervicothoracic region these changes cause shortening of the neck. The intervertebral discs may remain relatively normal, but sometimes are considerably thickened (Fairbank, 1949, Brailsford, 1952).

Little has been written concerning the neurologic changes caused by alterations in the configurations of the spinal canal incident to the architectural variations caused by Morquio's disease. Donath and Vogl (1925, 1927) indicated that the spinal cord itself was unaffected by the basic cartilaginous disorder which produced the vertebral changes. They stressed as a matter of practical importance the fact that the consequent shortness of the pedicles due to the premature synostosis of the centers of ossification of the body with those of the lamina led to a considerable narrowing of the entire spinal canal, either in the anteroposterior diameter or in all diameters. The space occupied by the spinal cord, therefore, was disproportionately large at the expense of the subarachnoid and the epidural spaces. This resulted in a corresponding narrowing of the spaces, and pressure upon the spinal cord. It is of interest that their communication referred to a condition designated by them as chondrodystrophic dwarfism, and possibly referred to achondroplasia. Nevertheless, it is more than likely that similar alterations may occur with Morquio's disease.

The coexistence of changes in the upper cervical spine and in the base of the skull in Morquio's disease was reported by Einhorn, Moore, and Rowntree (1946). They described a ten-year-old boy in whom the upper cervical vertebræ were fused, with anteroposterior compression of the spinal cord to the level of the fourth cervical segment. The base of his skull showed basilar invagination. Clinically this patient had developed loss of control of his fingers and lower extremities, and had been completely bedridden. Basilar impression was also reported by Feldman and Davenport (1951) and 2 of their other cases showed enlarged calvariums.

Rarely cases of osteochondrodystrophy are reported in which the spine is normal. Two such cases were reported by Shafar (1941).

Pelvic changes are an almost constant concomitant of chondroosteodystrophy. The iliac bones are flared outwards, and the true pelvis is contracted and deformed by the inward protrusion of the acetabulæ. The hip joints are deformed. The acetabular rims are thickened and the femoral heads fragmented, and sometimes actually destroyed. The femoral necks become thickened. With advancing years the joint deformities become even more pronounced. In some cases dislocation of the hips takes place. Changes in the joints of the extremities are somewhat less frequent, and as a rule no gross shortening takes place. The ends of the bones may become quite prominent because of enlarged epiphyseal structures. The joint spaces are widened. Later in life the deformities of the long bones may become less evident.

Osteochondrodystrophy may be confused with other bony dystrophies, including achondroplasia and dyschondroplasia. In typical cases the configuration of the vertebral bodies together with the changes in the extremities, particularly their articular ends, the disturbances in ossification of the epiphyseal lines and the familial history (which is not of much use because it also obtains in achondroplasia and dyschondroplasia) should provide some lead toward the correct diagnosis. The possibility of an atypical form of chondro-osseous defects was stressed by Jackson (1951) who reported 3 sibling dwarfs from a large and otherwise normal family with an unusual form of cartilaginous defect combined with club feet which bore some resemblance to the Morquio type. To some degree his patients also resembled cases of achondroplasia. He was of the opinion that the various osseous dystrophies represented irregular forms of osteochondrodystrophy which do not necessarily fall into rigidly defined groups.

Gargoylism (dysostosis multiplex) is considered another form of osteochondrodystrophy, but is attributed to a form of lipoidosis. It characteristically presents itself in patients with mental deficiency, a heavy and ugly facies, cloudy corneas and hepatosplenomegaly. The appearance of the vertebral column and ribs helps to differentiate gargoylism from osteochondrodystrophy, inasmuch as the platyspondyly and tongue-like configurations are not present with gargoylism, while the narrowed rib necks seen with gargoylism are not observed with other conditions. It might be mentioned that kyphoscoliosis with tongue-like projections from the anterior aspects of the vertebræ has been observed with the Ehlers-Danlos syndrome (cutis hyperelastica), but the clinical picture otherwise permits ready differentiation. Another rare condition in which chondrodysplasia occurs is chondroectodermal dysplasia (Ellis-van Creveld syndrome). In this condition the extremities are shortened, polydactyly and syndactyly appear, and there is hypoplasia of the nails, teeth and sometimes of the hair. The spine, however, is not affected.

A rare condition sometimes misdiagnosed as Hurler's disease or Morquio's disease is Léri's pleonosteosis, a genetically transmitted systemic disease involving mainly the connective tissue system with symptoms referable to the cutaneous, skeletal and ophthalmologic systems. In the cases described by Rukvina, Falls, Holt and Block (1959) the first in the American literature the vertebræ presented changes of considerable interest. The overgrowth of bone was manifested by increase in the heights of the pedicles and overgrowth of the vertebral bodies, suggesting in some ways the changes seen occasionally with acromegaly.

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DYSCHONDROPLASIA (OLLIER'S DISEASE).

Dyschondroplasia, or multiple enchondromata, is a non-hereditary developmental error characterized by bilateral multiple cartilaginous enchondroses which represent portions of epiphyseal plates which failed to develop into normal bone and had become incorporated into mature bone. These enchondromatous deposits are seen mainly in the long bones, and may cause expansion and consequent gross deformities. Occasionally exostoses are present. With the onset of puberty growth of the enchondromas terminates and calcification within the lesions results in an irregularly stippled radiolucent pattern. Dyschondroplasia with multiple hemangiomas is designated as Maffucci's syndrome. The disease rarely affects the vertebral column, but I have noted two instances in which small radiolucencies were observed in the transverse processes of a lumbar vertebra.

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HEREDITARY MULTIPLE EXOSTOSES (DIAPHYSIAL ACLASIS)

This is a hereditary disturbance in the nature of a mesodermal dysplasia. It is progressive from infancy to adolescence, and may advance until the growth period terminates. The condition affects principally the growing ends of bones, but may manifest itself in any bone formed in cartilage. It may become apparent during adolescence, when rapidly progressing growth may be accompanied by sudden progression in the size of some of the exostoses, making them apparent to the patient for the first time.

The exostoses may be unilateral or bilateral, and occur as single lesions or in multitudes. The distribution most of the time is asymmetric. The most common bones affected are the femurs and tibias, the upper portions of the humeri, the fibulas, the radius and ulna. Involvement of the pelvis, the clavicles, sternum, mandibles and ribs is less frequent. Exostoses projecting from the middle of the bone shafts are uncommon. Shortening of the growth of a long bone may produce visible deformity, such as Madelung's deformity of the forearm and wrist.

While infrequent, vertebral involvement with multiple exostoses has been recognized. A case with an exostosis within the spinal canal was reported by Ochsner and Rothstein (1907).

Involvement of the vertebræ with exostoses usually occurs in the neural arches. In themselves, these lesions are not of particular significance, but if they should be directed towards the spinal canal compression of the cord or nerve roots may follow. The vertebral bodies themselves are rarely affected. Slepian and Hamby (1951) found 19 cases in which neurologic complications were directly related to an encroaching osteochondral exostosis. Out of the 15 cases in which the sex was stated, 14 were males.

The spinal cord was involved in 10 cases. These authors reported 2 additional cases, 1 with a broad-based osteochondroma over the fifth and sixth cervical vertebræ, in which myelography revealed a complete block. Laminectomy showed the canal to be narrowed in the anteroposterior diameter, with elevation of the dura and displacement of the thecal sac to the right by a grumous bony growth.

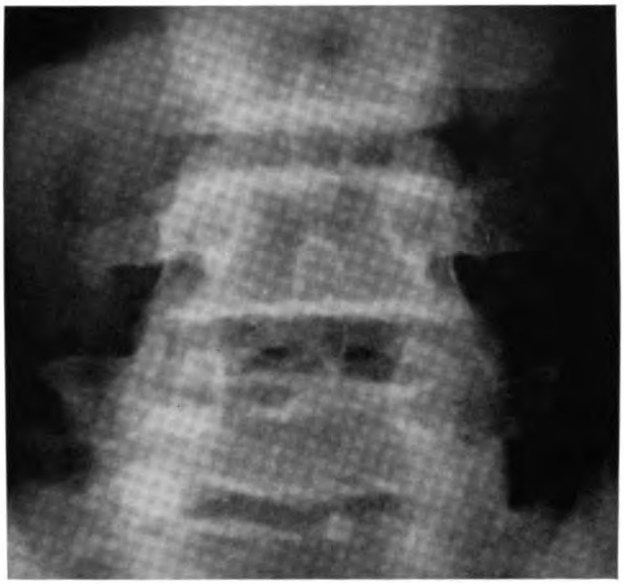

FIG. 97.—Small osteoma projects from the inferior aspect of the tip of the right transverse process of the fourth lumbar vertebra. Patient is a 37 year old man with hereditary multiple exostoses.

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ACHONDROPLASIA

This condition, which is associated with dwarfism, is also known as micromelia and chondrodystrophia fetalis. Although sporadic cases appear, it is of a hereditary and familial nature. It results from interference with endochondral ossification and failure of normal bone formation. The dwarfism is of the short limb type, associated with a large head and thick protruding lips. In many cases the hands present a so-called "trident" appearance, being broad and short with diverging fingers of almost equal length.

Achondroplasia is the commonest and most ancient known form of dwarfism. Females are affected somewhat more frequently. The intelligence of these individuals is often normal and not infrequently above average. The condition may be identified roentgenologically *in utero*, since failure of normal ossification of the long bone becomes apparent in the fetus at the end of the second month or early in the third month of gestation. Death *in utero* or premature birth may occur relatively frequently. Many achondroplastic infants die during the first year of life.

The dwarfism in these people is due mostly to lack of growth of the long bones. The histopathologic changes center chiefly about the epiphyseal plates. These appear broadened and jagged in outline, and present an irregular appearance which microscopically is represented by a haphazard and irregular arrangement of the chondrocytes. No reason is known for this disturbance in the cartilaginous pattern of the epiphyses. When growth ceases, the epiphyseal plate may disappear or be visible in a thin form for a long time.

Radiologic diagnoses of achondroplasia *in utero* are not at all infrequent. In 3 cases I had the opportunity of studying the fetuses both before and after delivery. The outstanding observation is a marked thinning of the osseous centrum of the vertebral bodies, with a disproportionately wide space between the vertebral bodies. These radiolucent spaces are due to unossified cartilage at the epiphyseal plates which radiologically are indistinguishable from the intervertebral discs. In addition, one can demonstrate not infrequently small tonguelike linear, projections from the anterior aspect of the ossified portions of the vertebral bodies. The narrowness of the vertebral bodies, diminished height and apparently increased length are quite characteristic of this condition. The 3 cases also presented a well-defined convexity of the lower dorsal and lumbar spine.

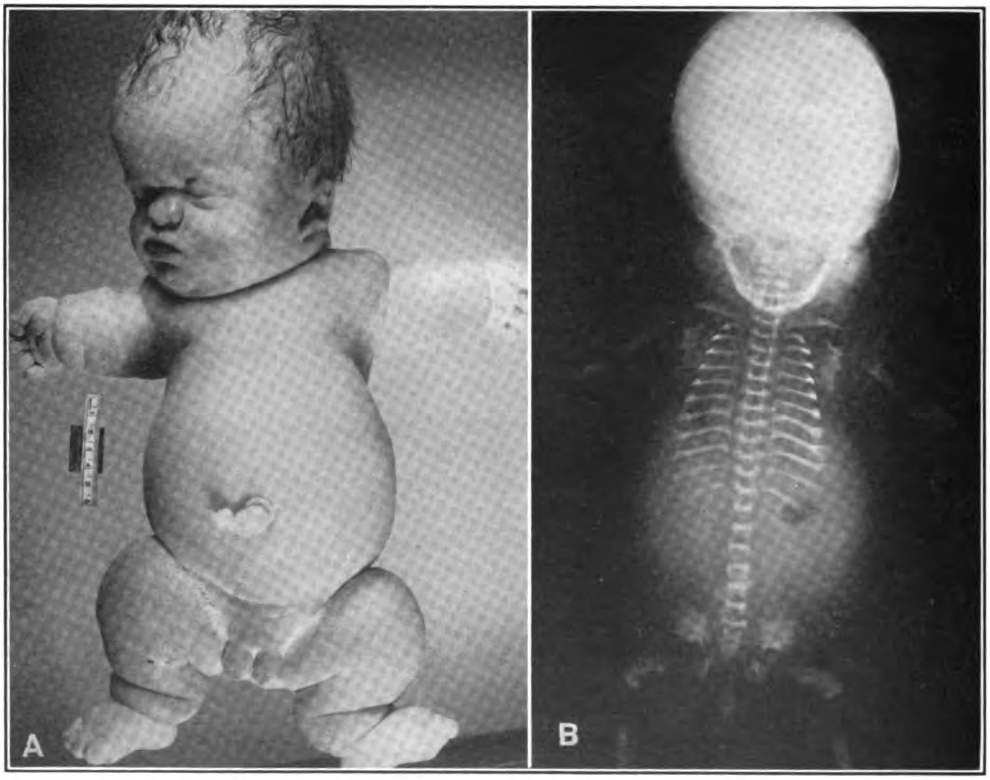

Fig. 98. Legend on opposite page.

Achondroplasia is often associated with broadening and shortening of the base of the skull, believed due to premature ossification and fusion of the bones about the foramen magnum. This may narrow the spinal canal and thus produce a variable degree in hydrocephalus by restriction of the circulation of cerebrospinal fluid.

In later life the vertebral bodies seem to recover their capacity for growth. As the patient approaches maturity the vertebral segments begin to assume a fairly normal appearance. In several cases observed by us anomalies of growth in the region of the fourth and fifth lumbar vertebræ were noted (Fig. 100). These structures are wedged between the iliac bones and are markedly sacralized. In addition the patient shown here presented evidence of basilar invagination, with diminution in the size of the odontoid process and the bodies of the first and second cervical vertebræ.

Changes in the alignment of the vertebral bodies are frequent, and an exaggerated lordosis is one of the more common changes. However the

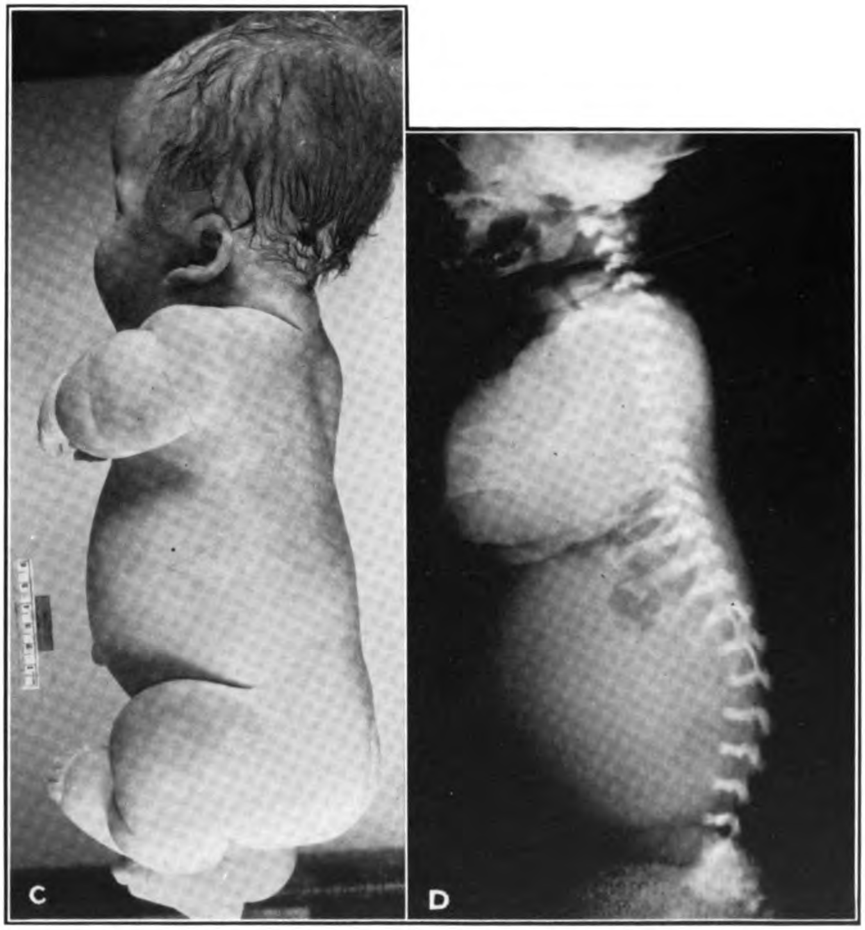

FIG. 98.—*A*, Photograph of stillborn female fetus with achondroplasia. *B*, Same case. Anteroposterior roentgenogram. Note the short limbs and small pelvic and shoulder girdle development, and the large head. The spine is straight. *C*, Same case. Photograph of infant in lateral position. *D*, Same case. Lateral roentgenogram. A dorsal curvature is present at the thoracolumbar junction. The vertebral bodies are small and the intervertebral discs are markedly enlarged. Deficient ossification of the pedicles and laminæ is present.

lordosis in many cases may be more apparent than real, being due to prominence of the buttocks. Kyphosis of the dorsal spine has been observed fairly frequently. The spinal canal in achondroplastic dwarfs is narrowed. It usually is of just sufficient diameter to accommodate the spinal cord, to the virtual exclusion of the subarachnoid and epidural spaces. The development of a dorsolumbar kyphosis in these individuals may therefore be associated with prominent neurologic symptoms, particularly if there should appear osteophytes which protrude into the lumen of the spinal canal. In such patients decompressive laminectomy may be required to provide relief.

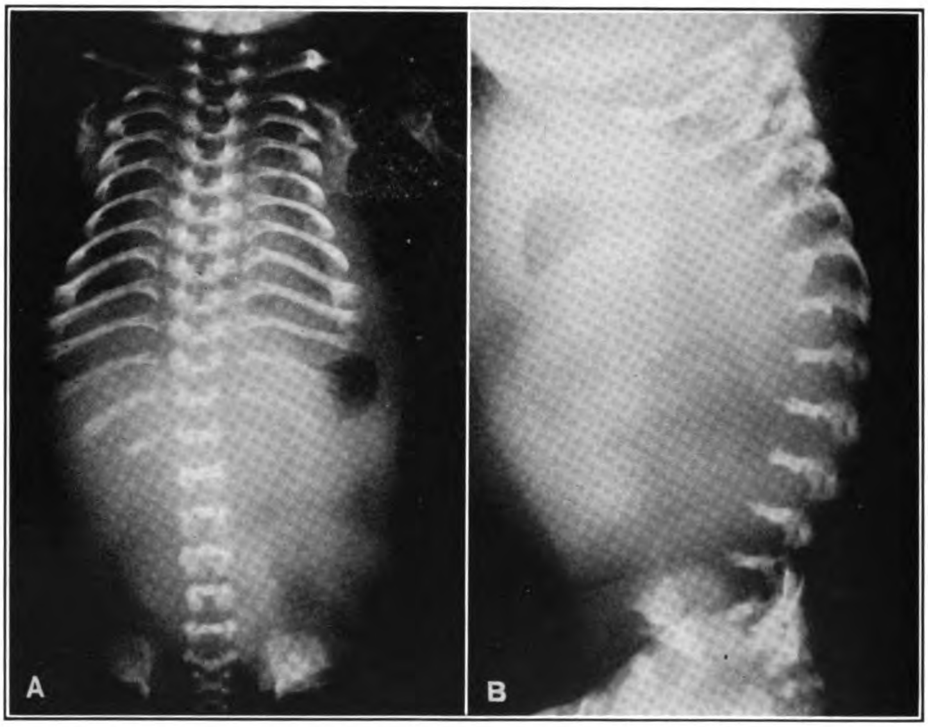

FIG. 99.—*A*, Anteroposterior roentgenogram of newborn infant with achondroplasia. Note ths widened intervertebral spaces and the poor development of the vertebræ. *B*, Same case as *A*, lateral view. The spine presents a thoracolumbar dorsad curvature, with small vertebral bodiee and markedly enlarged intervertebral discs. Note the tongue-like protrusions at the anterior aspects of the lumbar vertebræ. (See opposite page for Figure 99*C* and *D*.)

The development of a dorsolumbar kyphosis may sometimes proceed until a gibbus is formed. It is these individuals who are particularly predisposed towards neurologic complications. Vogl and Osborne (1949) suggested that in achondroplastic infants orthopedic measures be instituted before the child assumes the upright position to compensate for a dorsolumbar kyphosis before a gibbus forms.

Schreiber and Rosenthal (1952) reported 2 cases with partial paralysis of the legs and pain caused by herniated lumbar discs in achondroplastic dwarfs. They were operated upon with satisfactory results. These authors believed that whereas some achondroplastic dwarfs formed gibbi because of wedging of the dorsolumbar vertebral bodies, others may have a predis-

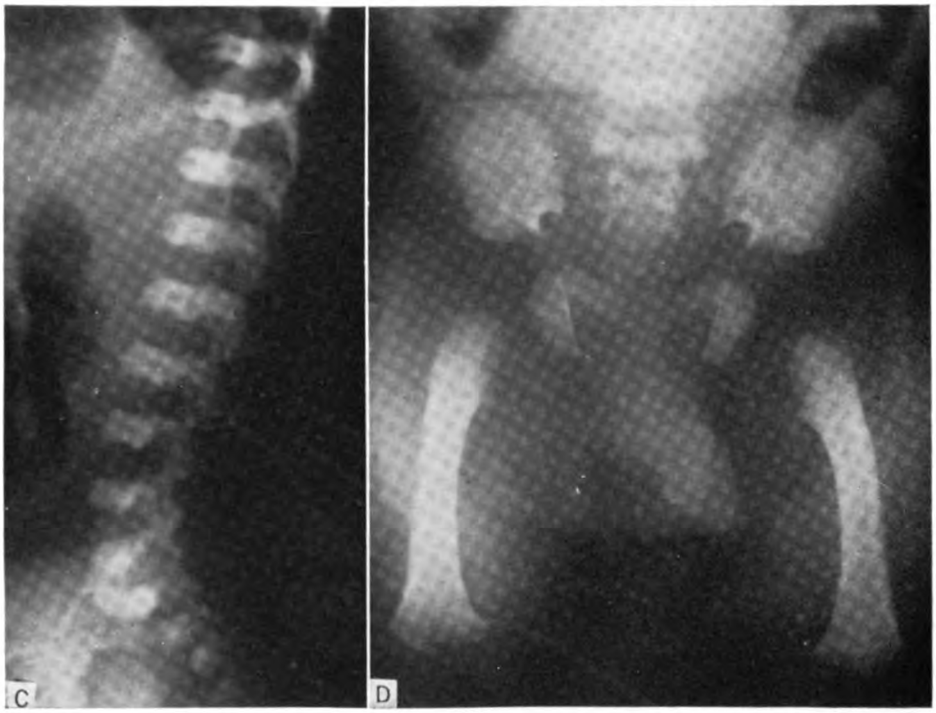

FIG. 99. *Continued.*—*C*, Another newborn infant with achondroplasia in a milder form. The vertebral bodies are larger than in the patient represented in *A* and *B*. The intervertebral spaces are wide. *D*, Same patient as *C*. The pelvis, hips and femurs show the characteristic osseous and joint changes.

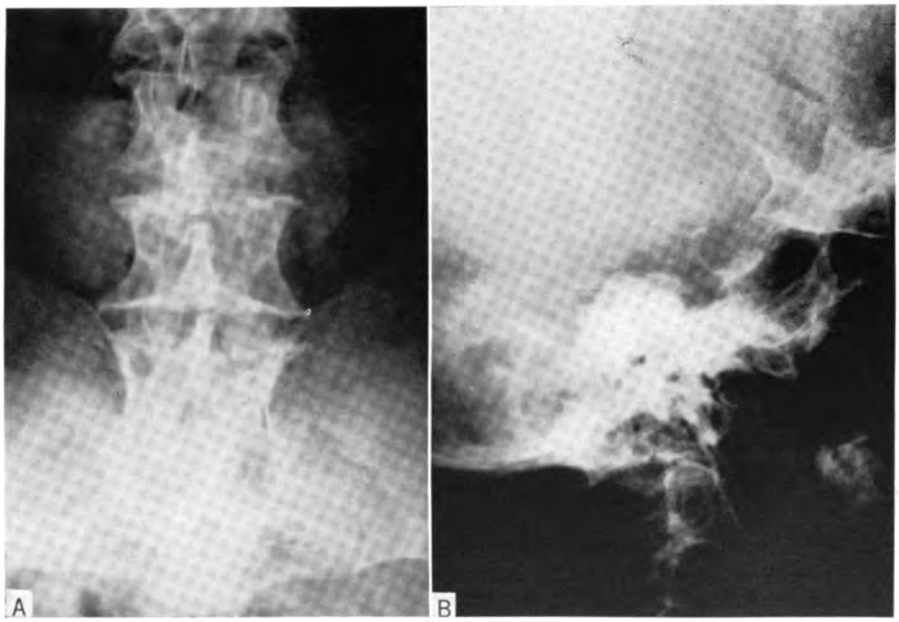

FIG. 100.—*A*, Lower lumbar spine in adult achondroplastic dwarf. The fifth lumbar vertebra is sunken between the iliac bones. The vertebræ are heavy, the pedicles are thick and close together, and the neural arches are also heavy. The inferior articular facets likewise are close together. *B*, The patient's cranium is large, but the base of the skull is shortened in the vicinity of the craniocervical junction. Moderate basilar invagination is present.

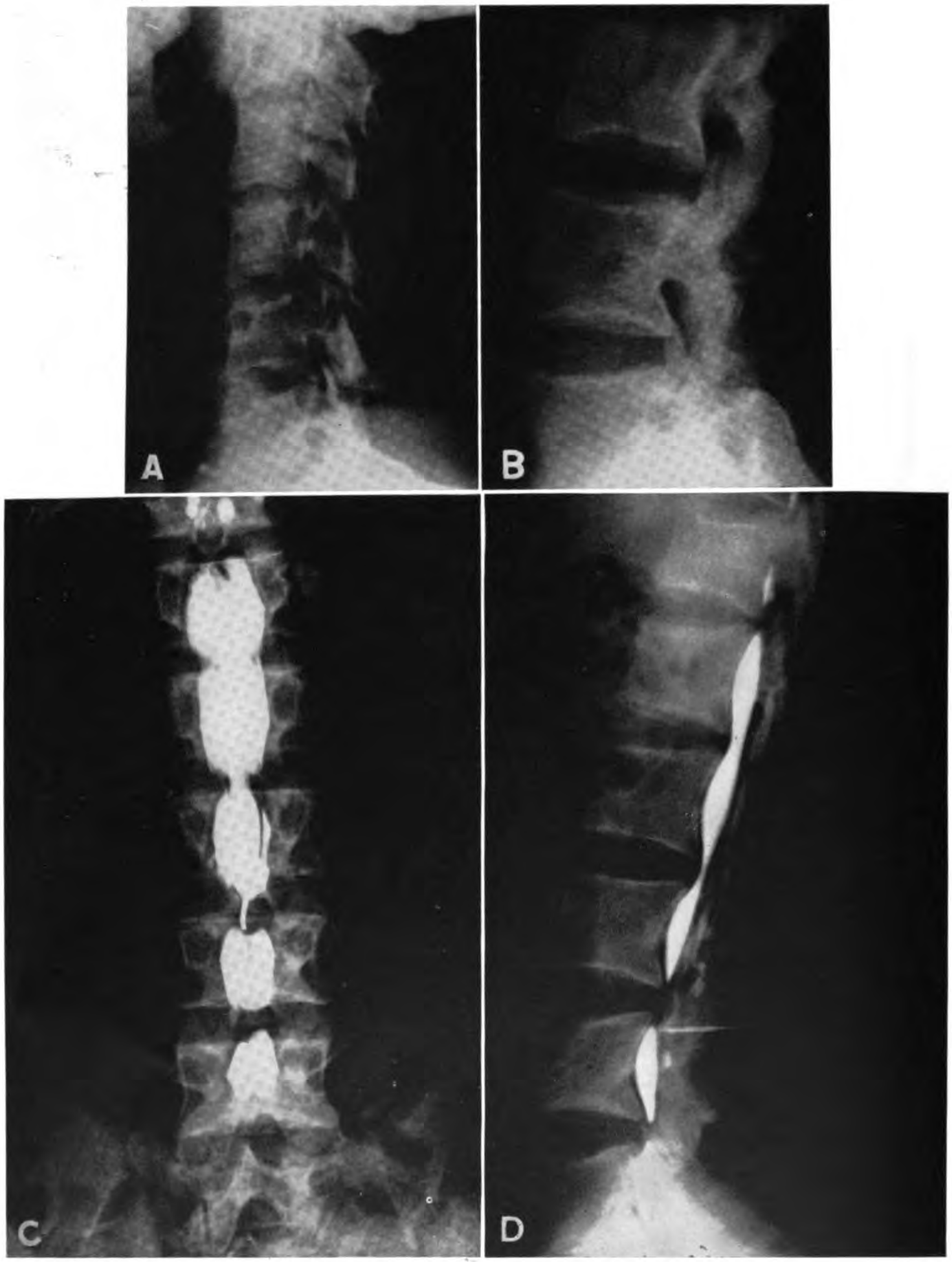

FIG. 101.—*A*, Oblique cervical roentgenogram of adult achondroplastic dwarf. Note the elongation and marked narrowing of the intervertebral foramina. *B*, Same patient, demonstrating similar malformation of the lumbar intervertebral foramina. *C*, Because of intense radicular symptoms due to cauda equina pressure, myelography was done. Note the wedging of the fifth lumbar vertebra between the iliac bones. Transverse defects are present at each discal level. *D*, Lateral myelogram reveals a thin pantopaque column due to narrowing of the spinal canal. The bulging discs together with the heavy neural arches, hypertrophied ligamenta flava and narrowing of the spinal canal, produced severe cauda equina pressure. Relief of symptoms followed multiple laminectomies. (Courtesy of Dr. Joseph A. Epstein.)

position to herniated discs, probably attributable to the inherent weakness of the spinal column. Myelographic examination in their patients showed a complete block at the interspace between the second and third lumbar interspaces in both cases. They noted at operation that the spinal canal of both these people were narrowed and shallower than normal. Neither of the two had changes in alignment of the vertebral segments, nor were there any evidences of osteophytes encountered at operation. Previously Albrecht and Ramzi (1926) had reported a case of compressive myelitis in an achondroplastic dwarf in whom the symptoms were relieved by laminectomy. Freund (1933) had also reported on the occurrence of spastic paraplegia in achondroplasia due to the projection into the spinal canal of osteophytic overgrowth encroaching upon the narrowed spinal canal. Spillane (1952) reported 3 cases with neurologic complications. Two had severe cauda equina compression due to multiple massive intervertebral disc protrusions, and 1 had a spastic tetraplegia due to subluxation of the second on the third cervical vertebræ.

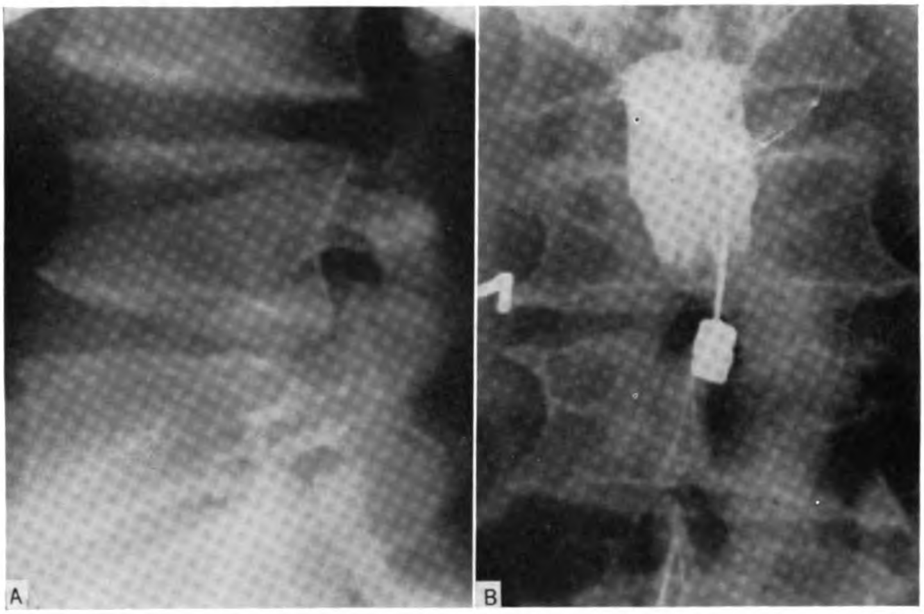

FIG. 102.—*A*, Adult male achondroplastic dwarf with intense radicular pain. The intervertebral foramina as seen on the lateral film are elongated and narrowed. The pedicles and laminæ are heavy. The vertebral bodies are flattened. *B*, Myelogram shows a complete block at the third lumbar interspace. A relatively small herniated disc was found at operation. The canal was small, so that even a small intrusion caused a complete block.

Recently 2 adult patients with mild achondroplasia presented themselves with neurologic deficits and back pain indicative of spinal cord and cauda equina pressure. One had a protruded nucleus pulposus completely blocking the spinal canal at the second and third lumbar interspace. The other, (Fig. 101), had multiple levels of compression from the tenth dorsal to the fifth lumbar interspace due to dorsal bulging of the intervertebral fibrocartilages, compressing the cord and cauda equina against the neural arches. Both patients responded well to surgical relief of the cord pressure.

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OSTEOGENESIS IMPERFECTA

Osteogenesis imperfecta is a fairly common hereditary disturbance of normal mesenchymal development which produces a marked alteration in the skeletal structure characterized by softening and brittleness in varying proportions.

Other names which have been applied to this condition are idiopathic fragilitas ossium, osteopsathyrosis, brittle bones with blue scleræ, osteosclerosis, osteitis parenchymatosa chronica, dystrophia periostalsis, periosteal aplasia and periosteal dysplasia. Brailsford suggested that all these terms be dropped, and the term osteogenesis imperfecta be extended to include fetal, infantile, adolescent and adult types (1943). Chont had also suggested in 1941 a revision of nomenclature to include two forms of the disease: osteoporosis imperfecta congenita, all cases of which either die *in utero* or are born with the disease, constituting thereby a fetal and infantile variety. The second major classification would be osteogenesis imperfecta tarda occurring in infants apparently normal at birth and in whom the disease occurs early in life, usually in the second year. This group could be subdivided into a hereditary group with blue scleræ and non-hereditary group which may or may not have blue scleræ. Caniggia, Stuart and Guideri (1958) regard as valid the unitary concept of fetal and adult forms of osteogenesis imperfecta in the genetic, pathogenetic and histopathological domains. They recommend that the term fragilitas ossium hereditaria, Typus Vrolick, or osteogenesis imperfecta Vrolick type, or briefly Vrolick's disease be used to indicate the congenital fetal form of the disease. The adult type can be referred to as osteogenesis imperfecta Ekman-Lobstein type, or Ekman-Lobstein disease.

Fairbank (1948) in a report of 12 cases of osteogenesis imperfecta mentioned 3 types, the first being the thick bone type with severe bony changes particularly in the ribs, together with stunted limbs such as may be seen with achondroplasia. The second type presented slender fragile bones with osteoporosis of the entire skeleton. In this type the vertebral bodies may be translucent, shallow, spread and biconcave with biconvex intervertebral discs. A third type, which he mentioned as very rare, presents pronounced honeycombing of the bones of a progressive nature. He termed this osteogenesis imperfecta cystica. Rosenbaum (1944) questioned whether there was any difference in the type of osteogenesis imperfecta in which intra-uterine fractures occurred or in which only post-natal fractures occurred. He reported cases of the 2 types in the same family, tending to disprove the duality of the syndromes. There does not appear to be sufficient evidence to warrant the division of this disease into 2 specific categories. The evidence appears to point towards the existence of several forms of the same disease, some of which become manifest during intrauterine life; others of which appear to be milder in the extent of osseous change. The prognosis for those individuals in whom the disease is manifest early in life is not as good as those who present later development of osteogenesis imperfecta.

Clinically there are marked variations in the severity of the disease. The usual clinical picture is one of a short, poorly-nourished individual who presents many bony deformities associated with multiple fractures, some of which appear practically spontaneously. The skull tends to be triangular in shape, and dental development is retarded. Excessive mobility of the

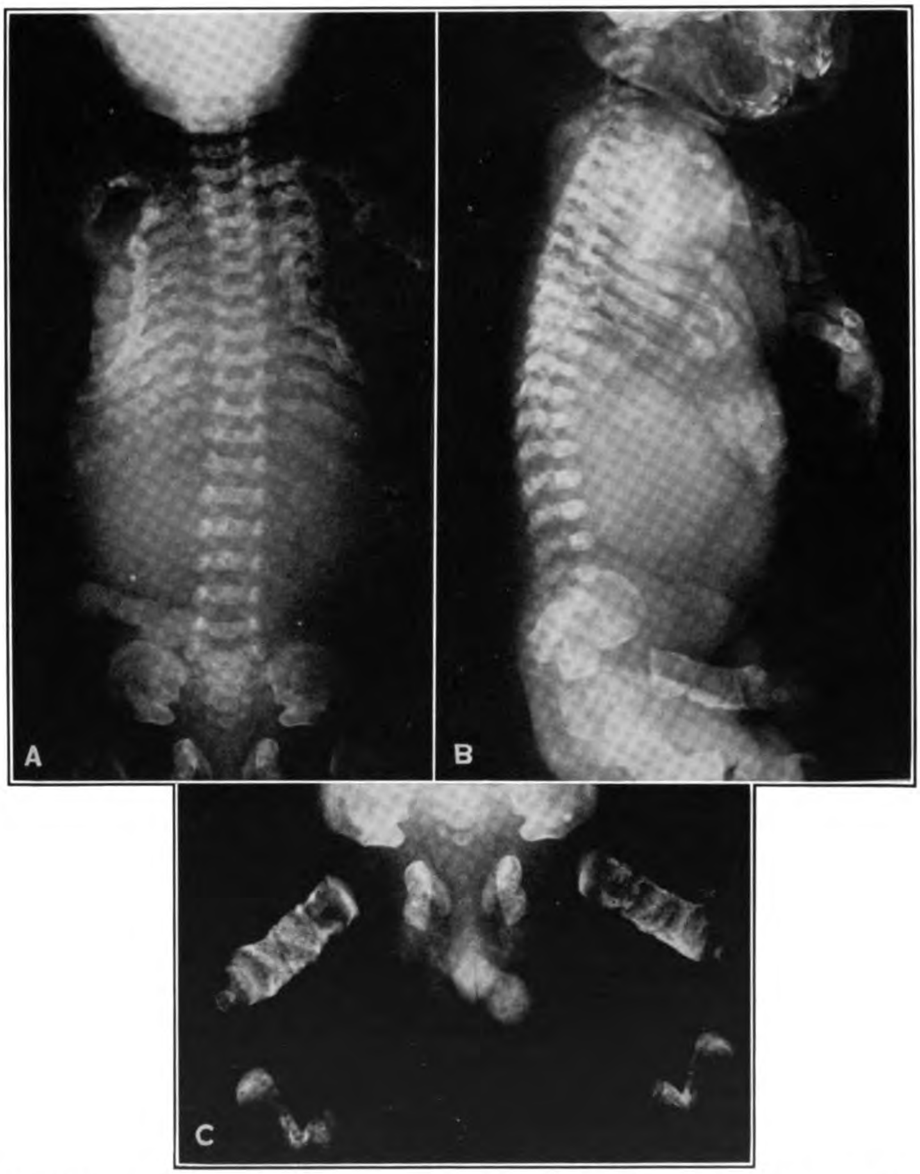

FIG. 103.—*A*, Anteroposterior roentgenogram of stillborn infant with osteogenesis imperfecta. Note the wrinkled appearance of the ribs, and the inward collapse of the supradiaphragmatic costal structures. The vertebral column is quite straight. *B*, Same case, lateral view. Note the dorsal convexity at the thoracolumbar junction, and the anterior wedging of the lower thoracic and upper lumbar vertebræ. Ossification appears adequate. *C*, Same case, illustrating the marked deformity of the lower extremities.

joints may be demonstrated, due to relaxation of the articular capsules and restraining ligaments together with poor muscular development. The presence of blue scleræ is considered requisite for the hereditary type of the disease, but this may not always be present. Deafness is a fairly frequent accompaniment, and is attributed to ankylosis of the ossicles, or in more advanced cases, to actual involvement of the bones of the middle ear.

The pathologic changes associated with osteogenesis imperfecta are imperfect formation and calcification of the bony trabeculæ. Islands of cartilage are seen, especially beneath the periosteum, in which imperfectly calcified osteoid trabeculæ are present, which replace normal bone. The periosteum may be thickened, beneath which the bone is discontinous and fragmentary. The medullary content is in part fibroid, lymphoid or fatty. Studies by Fairbank (1948) indicated that there may be a great variation in the development of the osteoblasts. He pointed out that while a deficiency in the number of osteoblasts had been stressed, his studies showed that the number may be quite numerous although the formation of bone was deficient. Knaggs (1926) had mentioned before that a deficiency in osteoblasts was common, and that formation of cartilage cells from the periosteum instead of the osteoblast is the most characteristic pathologic change.

Prepartum diagnosis of osteogenesis imperfecta occasionally can be made from abdominal roentgenograms. The limbs of the fetus present a slender appearance, and fractures sometimes can be identified. In others the wrinkled appearance of the bones, particularly of the ribs, is quite diagnostic.

Roentgenologic examination of the extremities in cases with intrauterine or fetal osteogenesis imperfecta may reveal a marked crinkled appearance of the long bones. These are short and relatively stumpy, with practically no distinguishing markings between the cortex and medullary canal. Some subperiosteal thickening may be present, giving rise to irregular, rather scalloped peripheries of the tubular bones. These may also show numerous radiopaque bands due to the bending over of the softened bone. Deformities often are marked, some of the bones presenting almost right angle configurations in the middle of their shafts, others appearing to be collapsed upon themselves in an irregular accordion-like fashion. The ribs may be markedly involved and the thoracic cavity greatly compressed because of the marked softening and irregularities of the ribs. It is interesting to note that this change is most marked in the portion of the thoracic cage occupied by the lungs. The infradiaphragmatic ribs usually are kept more or less in position by the underlying liver, spleen and other abdominal viscera. Other cases of osteogenesis imperfecta of the intra-uterine and fetal type present changes which are much less marked. In such instances the fracture lines are sharply visible and there may be considerable displacement of the bony fragments. Angular deformities of the long bones, particularly the femurs and legs, are frequent. The degree of deformity of the ribs may be less marked, and rather than presenting an irregular crenated type of bony deformity the ribs may be thin and slender. As maturity is reached the disease may take one of two roads. In some individuals the development of bone slowly proceeds in a more or less normal fashion so that by the time adolescence is reached and growth stops, the bony structures are deformed, thin, but still capable of carrying the weight of the child. In such instances further growth is accompanied by less and less deformity, and these individuals may reach maturity and be symptomless. It is even possible for some of these patients to be afflicted with osteogenesis imperfecta and go through life without the diagnosis being definitely established, inasmuch as symptoms are so few as to make medical attention unnecessary in the early stages of life when the disease might have been more easily recognized. Other individuals may show marked deficiency in development of bone, and rather than go on to healing the bones may present a tubular appearance. The cortex and medullary canals are relatively well differentiated but the bones are very soft and deformities and fractures occur frequently.

The appearance of the vertebral column in patients afflicted with osteogenesis imperfecta is variable. Spinal curvatures are frequent. The vertebræ become softened, with resultant compression and widening of the bodies and ballooning of the intervertebral discs. In the more advanced cases compressive action on the spine produces pronounced flattening of the vertebræ which assume the appearance of a biconvex flat lens. It has been noted that occasionally the vertebral bodies at the lower lumbar levels are somewhat increased in height in patients who have the greatest extent of spinal compressive change. The vertebræ are demineralized, the compact bone being considerably thinned and the cancellous bone reduced in volume so that the trabecular spaces are widened. The remaining trabeculæ, however, are quite well calcified.

Observations in a case of intra-uterine osteogenesis imperfecta were made by post-mortem roentgenograms of the skeleton immediately after the delivery of the stillborn fetus, and in these the vertebral column was of particular interest (Fig. 103). The alignment of the vertebral bodies was about normal for the fetus at term, insofar as the cervical and thoracic spine was concerned. The lumbosacral curve was slightly more developed than one might have anticipated. In the anteroposterior view, the vertebral column was quite straight. In the cervical spine the middle cervical segments presented a definite anterior wedging of the vertebral bodies while the posterior aspect of these vertebral bodies maintained their height quite well. The dorsal spine showed a definite decrease in the height of the vertebral bodies together with a proportionate increase in the width of the intervertebral spaces. The superior and inferior aspects of the respective vertebral bodies were quite straight and parallel to each other. The thoracolumbar junction was of interest in that there was a rather abrupt wedging of the anterior aspect of the bodies of the eleventh and twelfth dorsal vertebræ, and to a lesser degree of the anterior aspect of the body of the first lumbar vertebra. The other lumbar vertebræ were rather small but otherwise well formed. The anterior and posterior venous channels entering the vertebral bodies were conspicuous by their absence in all of the vertebræ. The neural arches presented the same type of crinkled appearance as was noted in the markedly altered long bones. However, the interpedicular spaces were not visibly disturbed and the pedicles presented a firm bony appearance.

Another case of osteogenesis imperfecta in a newborn infant presented a considerably different appearance of the vertebral bodies. In this patient the bony structures did not show the accordion-pleated wrinkling noted in the previous case. The medullary canals were fairly well identified and the fracture lines in the arms and legs were relatively sharply demarcated. In this patient a definite narrowing of the bodies of the dorsal vertebræ was noted. The intervertebral spaces were markedly increased in height. In this patient the vertebral column was well aligned in the anteroposterior view, while in the lateral view a slight but fairly definite tendency towards a roundness dorsally in the thoracolumbar region could be demonstrated. The degree of thinness of the vertebral bodies was greatest in the thoracic spine, while in the lumbar spine the vertebral bodies returned to an almost normal height. The vascular indentations of the anterior and dorsal aspects of the vertebral bodies were inconspicuous but could be faintly made out in the lumbar segments.

With advancing growth the height of the vertebral bodies increases and their bony density improves. However, in certain instances in which the disease is markedly advanced, the degree of demineralization of the vertebral bodies is so pronounced that the structures are virtually indistinguishable on the roentgenogram. In such cases the vertebral bodies become reduced to only a fraction of their normal height and assume biconvex deformities associated with enlargement and biconcave configurations of the intervertebral discs. Continued observation of such a patient showed a marked improvement in the density of the flattened and deformed vertebral bodies over a period of years, so that when the condition was apparently healed the deformity of the vertebral bodies was quite marked, although the bony densities had returned to a fairly normal state. In this case the vertebræ presented marked broadening and flattening, with increased densities of the margins of the vertebral bodies. There also was an associated spreading of the interpedicular distances both in the thoracic and lumbar segments, considered part of the picture of spreading of the vertebral bodies associated with the softening accompanying osteogenesis imperfecta.

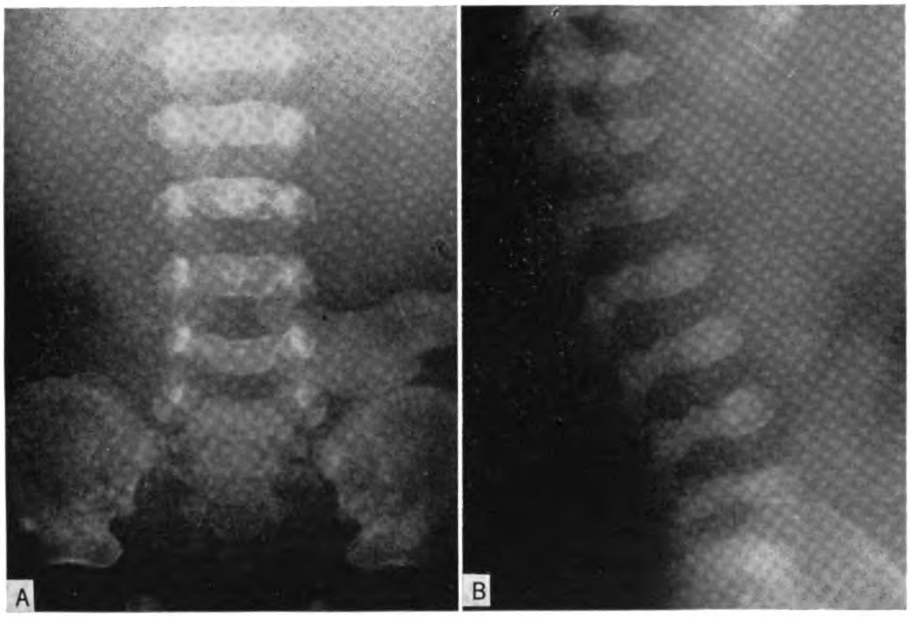

FIG. 104.—*A*, New born infant with osteogenesis imperfecta. The vertebræ are demineralized. Note the area of increased density in the center of the vertebral bodies. *B*, Lateral view of thoracolumbar spine. The lower thoracic vertebræ present a tongue-like anterior aspect. The lumbar vertebræ are biconvex. Note the increase in the density in the central areas.

Osteogenesis imperfecta with a relatively latent clinical course may be associated with a less pronounced degree of biconvex deformity of the vertebral bodies. In such cases, the length of the pedicles of the lumbar vertebræ as seen in the lateral view appeared to be increased, with a consequent apparent widening of the spinal canal. It is worth mentioning that in 1 case the child's mother was known to have had osteogenesis imperfecta, and that roentgenologic examination of her spine showed no deformity.

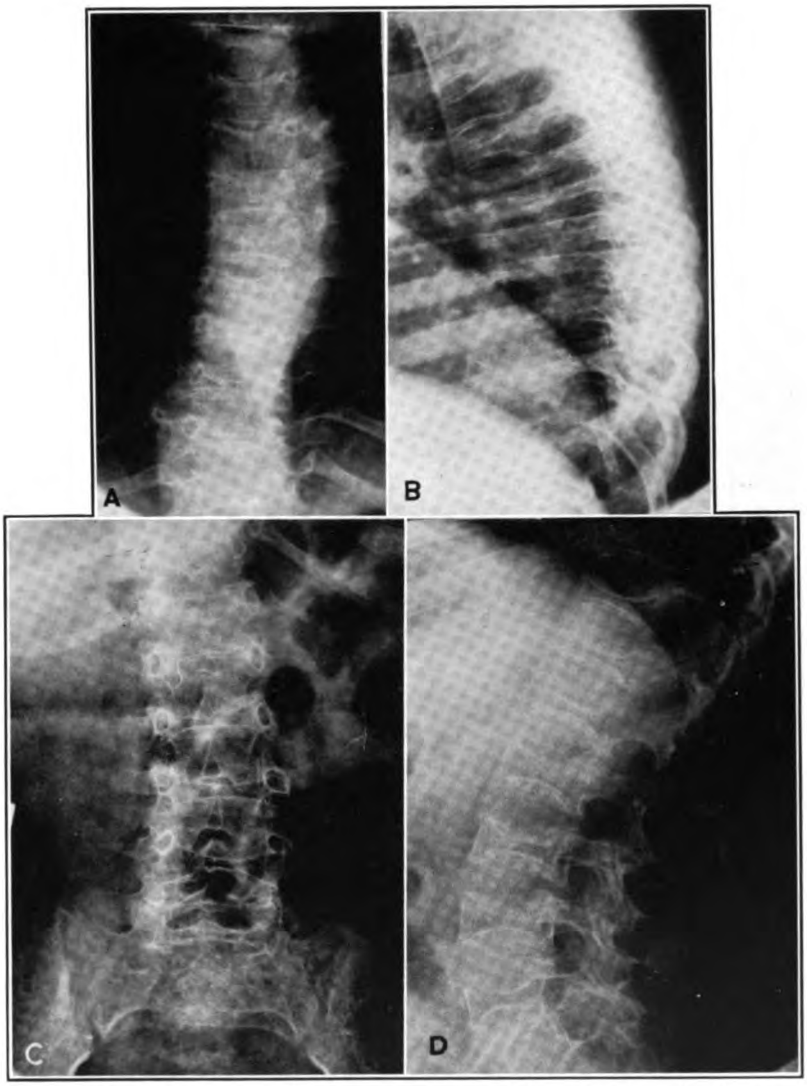

FIG. 105.—*A* and *B*. Anteroposterior and lateral view of thoracic spine of a nine-year-old girl with osteogenesis imperfecta. The vertebral bodies are markedly flattened, and the intervertebral discs are widened and bulging. *C* and *D*, Same case as *A* and *B*. Anteroposterior and lateral views of the lumbar spine. The vertebral bodies are biconcave, with marked bulging of the intervertebral discs. Note the peripheral condensation outlining the vertebral bodies.

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OSTEOPETROSIS

Osteopetrosis is also known as Albers-Schönberg's disease, marble bones, osteosclerosis fragilis generalisata and osteopetrosis generalisata. The disease is of familial nature and has been encounted in all decades of life from intra-uterine life to the eighth decade. The cause for the condition is unknown. It affects both sexes equally, and its severity may vary markedly from patient to patient. In some, particularly those with the disease *in utero* or during the first year of life, the progression is rapid, with complete involvement of the entire skeletal system, anemia, optic atrophy retarded growth and fatal outcome within a relatively short time. Other cases present a more attenuated form and the diagnosis may be established in the course of roentgenologic examinations made for other purposes. The disease is characterized to some degree by activity and remissions, or there may be complete cessation of the developmental error. Schulte (1951) divided Albers-Schönberg disease into three groups, including a malignant form of osteosclerosis in children with brittle bones and anemia, a benign form without anemia but bone sclerosis and weakness, and osteosclerosis with neither fractures nor anemia.

The anemia associated with osteopetrosis is ascribed to myelophthisis caused by encroachment on the marrow cavity. Metaplasia of extra-medullary hematopoietic centers accounts for enlargement of the liver, spleen and lymph nodes. With extensive skull involvement constriction of the foramina such as the optic foramina and the auditory canals produces symptoms referable to the various cranial nerves affected. However, it is remarkable that even when the skull is extensively altered relatively few cranial nerve deficiencies are noted in some patients. In spite of the apparent hardness of the bones, spontaneous fractures are not uncommon, and cases sometimes are first observed as a result of fractures produced by slight trauma.

Vertebræ affected with osteopetrosis are described as marble gray on section. The bodies are either uniformly dense or present as two heavy sclerotic bands on the upper and lower aspects of the bodies with a lucent zone between. The channels for the anterior and posterior veins may be exaggerated. The bony architecture is either irregular or distorted. Osteoblasts are normal or increased while osteoclasts are decreased or absent. According to current concepts osteopetrosis cannot be explained purely on the basis of diminished osteoclastic activity. Enticknap (1954) observed that both osteoblastic and osteoclastic activity in his patient with osteopetrosis were undiminished. Engfeldt and his coworkers (1954) demonstrated that normal bone resorbed by osteoclastic activity is replaced by immature bone which does not progress to normal bone.

Radiographic examination of the vertebral column in infants with osteopetrosis frequently discloses a marked increase in the density of the upper and lower plates, with a relatively lucent zone between. The notching of the vascular channels in the anterior and posterior aspects of the vertebral bodies often is accentuated, and the neurocentral synchondroses assume a broadened, flattened appearance. The neural arches are very dense but not malformed, and the appearance of the ribs is equally striking. No definite change in the alignment of the spinal column occurs.

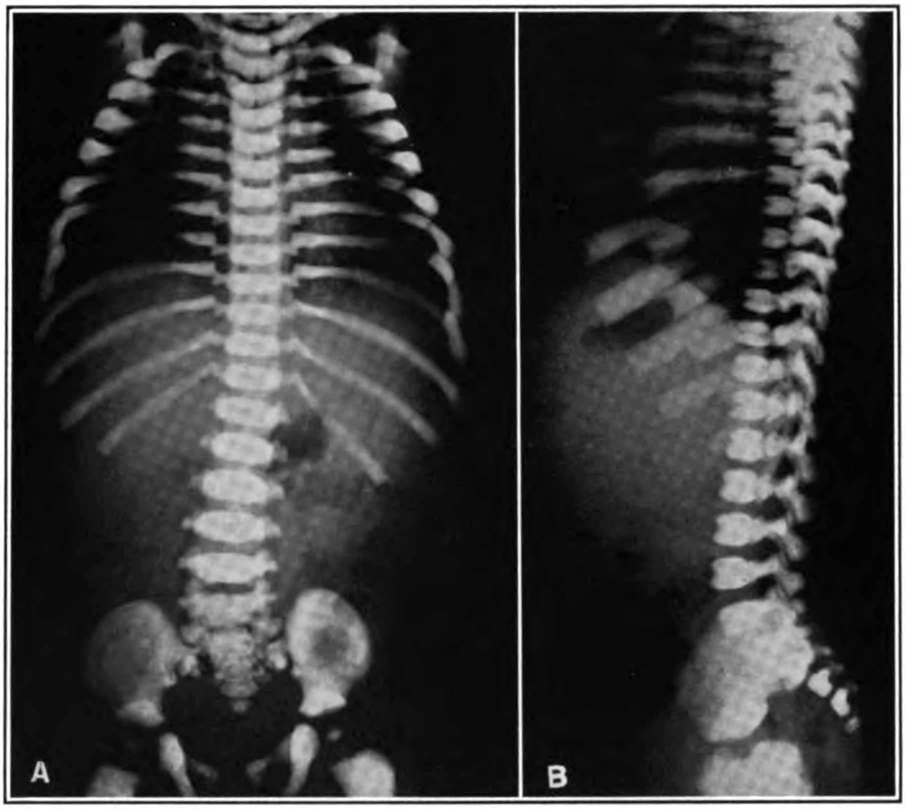

FIG. 106.—Anteroposterior and lateral roentgenograms of infant with osteopetrosis.

One of our patients with osteopetrosis was observed from birth to the age of three years. The condition was inexorably progressive. The child became blind, but was of normal intelligence and possessed acute perception of what was going on about her. The skeletal growth was retarded, the chest small and the abdomen markedly protruberant because of pronounced hepatosplenomegaly. Her vertebral column remained well aligned, but growth of the individual vertebræ was slow, with no gross distortion of configuration. The increased density of the upper and lower plates was striking, and the V-shaped notches on the anterior aspect of the bodies was persistent.

Adults with osteopetrosis likewise present a "sandwich" appearance of the vertebral bodies, with a lucent zone between. In some the sclerosis also affects the posterior aspect of the body producing a somewhat "C" shaped appearance.

While the radiologic picture of osteopetrosis is one of great density, the true nature of the osseous structures does not conform with this impression of hardness. The bone itself varies from a granite-like density to a consistency like that of chalk. Striking variations in density occur in various areas in the same patient. In adults it is not infrequent to discover the disease incidentally, an observation which emphasizes the difference between the early and late manifestations of the disease.

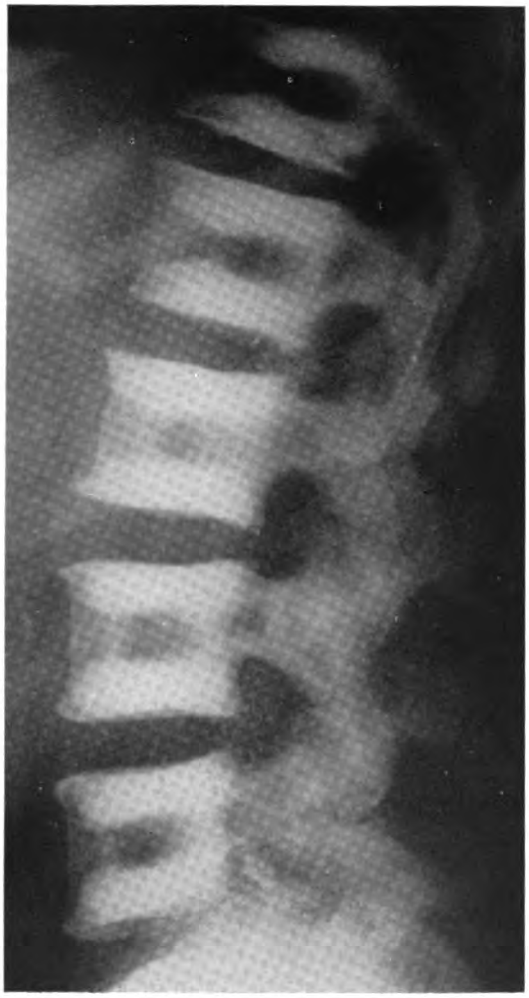

Fig. 107.—Osteopetrosis in an adult. Note the marked density in the upper and lower aspects of the respective vertebræ producing a "sandwich" appearance. (From Hinkel and Beiler, Am. J. Roentgenol., courtesy of Charles C Thomas, Publisher.)

The bony structures of the remainder of the skeleton in osteopetrosis is characterized by a peculiar tubular overgrowth of bone with practically complete obliteration of the medullary canal in advanced cases. The bones are also characterized by a peculiar cork-like configuration of the metaphyseal ends extending upwards into the diaphyses of the bones. On overexposed films of the long bones varying areas of radiolucency of a thin linear nature immediately beneath the periosteum may be outlined, giving the appearance of "bone within bone." These correspond to areas of fibro-osseous metaplasia.

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Fibrous Dyplasia (Fibrocystic Disease of Bone)

Although brought into prominence by the report of Albright, Butler, Hampton and Smith (1937) as a syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation and endocrine dysfunction with precocious puberty in females, this condition has been recognized as a separate entity for a considerably longer time. Albright and Reifenstein (1948) mentioned that in a review of von Recklinghausen's original monograph (1891) a number of conditions which certainly were not hyperparathyroidism were encountered. They believed that 2 out of the 3 famous cases in the monograph which had been considered examples of hyperparathyroidism apparently reflected the syndrome described by Albright and his co-workers. Albright commented that "as far as the bone manifestations of the syndrome are concerned, von Recklinghausen preceded the author and his colleagues by forty-two years." Albright and Reifenstein agreed with Jaffe and Lichtenstein that the term best applicable to this condition is simply fibrous dysplasia, omitting the word polyostotic in order to include cases confined to one bony lesion.

In its completely developed form, fibrous dysplasia affects multiple bones which have a marked tendency to be unilateral in their distribution, although both sides of the body may be affected. The patient presents brown, unelevated pigmented areas of the skin which are usually on the same side of the body as the bony lesion. This is accompanied in females with an endocrine dysfunction characterized by precocious puberty. The most common grouping of bone involvement is one leg associated with lesions in the pelvis and skull together with scattered lesions in the metatarsal and phalangeal bones. The femurs and humeri also frequently are affected as well as the small bones of the hand. As a rule the proximal portions are more affected. The pelvis, tarsal and carpal bones, the sternum and mandible are affected less frequently and the vertebræ are seldom involved. The affected bones present considerable widening of the medullary canal with thinned cortices. The medullary canal may be attacked by a thick white rubbery fibrous tissue which contains zones of scattered hyalin cartilage. The normal bony architecture of both the cortex and the medulla becomes disrupted by a dense fibrous tissue reaction with many fusiform spindle cells containing pale staining nuclei and a vague cystoplasmic outline. The fibrous tissue may contain small trabeculæ of primitive metaplastic bone. Occasional small cysts containing amber fluid may be present. The transition from pathologic to normal bone is often abrupt. The abnormal tissue has a gritty character due to the presence of the spicules of primitive bone (Fairbank, 1950; Russel and Chandler, 1950). Laboratory findings are usually of little help. The blood calcium and phosphorus usually are within normal limits. The phosphatase value varies considerably, and in some cases is increased. No single study, be it clinical, radiologic, operative or pathologic can sharply differentiate fibrous dysplasia from bone cysts or giant cell tumors.

The onset of this condition is usually insidious. First indications may be brought about by a fracture following a slight trauma or by a fairly rapidly appearing deformity of an extremity secondary to a local enlargement of the bone. A considerable asymmetry of the face or body also appears later in the disease. Pain and stiffness are less common initial symptoms. The skull changes of fibrous dysplasia may progress to the point where the disease assumes the proportions of leontiasis ossia with gross deformity of the face.

Fibrous dysplasia is regarded by Lichtenstein and Jaffe (1942) as a congenital developmental anomaly. Schlumberger (1946) was of the opinion that this was a true dystrophy in which the tendency of the mesenchyme to normal ossification is blocked or prevented by a fundamental local disturbance yet to be demonstrated. Albright and Reifenstein held that there probably is an endocrine disturbance related to a neurologic or embryologic defect disturbing the afferent impulses to the anterior lobe of the pituitary gland. Neller (1941) believed that polyostotic fibrous dysplasia was due to multiple embryonic defects, stating that the simultaneous involvement of many different regions was evidence against a defect in a single system. The coexistence of other congenital defects with fibrous dysplasia has been reported by Stauffer, Arbuckle and Aegerter (1941) and by Coleman (1939). It would appear that up to the present time no one theory has accounted for the multiple manifestations of fibrous dysplasia of bone as associated with cutaneous and endocrine disturbances.

The radiologic changes in the long bones reflects the widening of the medullary canals and thinning of the cortex. Single or multilocular cysts occur together with various deformities incident to fractures and bending, changes induced by the softening of the bone. In some instances a characteristic ground glass appearance of the medullary cavity is observed, a change apparent more often in younger patients. In older people the cystic changes may become more pronounced, grotesquely deforming the bones and producing multiloculated cystic areas. Changes in the skull may simulate those of Paget's disease, and sometimes the two conditions are misdiagnosed one for the other. This situation can be avoided by due attention to the more characteristic manifestations of Paget's disease in the long bones, pelvis and spine.

Involvement of the vertebral column is rare. A review of the literature indicates only sporadic examples of such change. Albright and Reifenstein (1948) mentioned a case in which the entire spine was involved, while in another only the cervical vertebræ were affected. Spinal involvement was observed in two cases reported by Furst and Shapiro (1943), and in two of the cases reported by Dockerty, Ghormley, Kennedy and Pugh (1945).

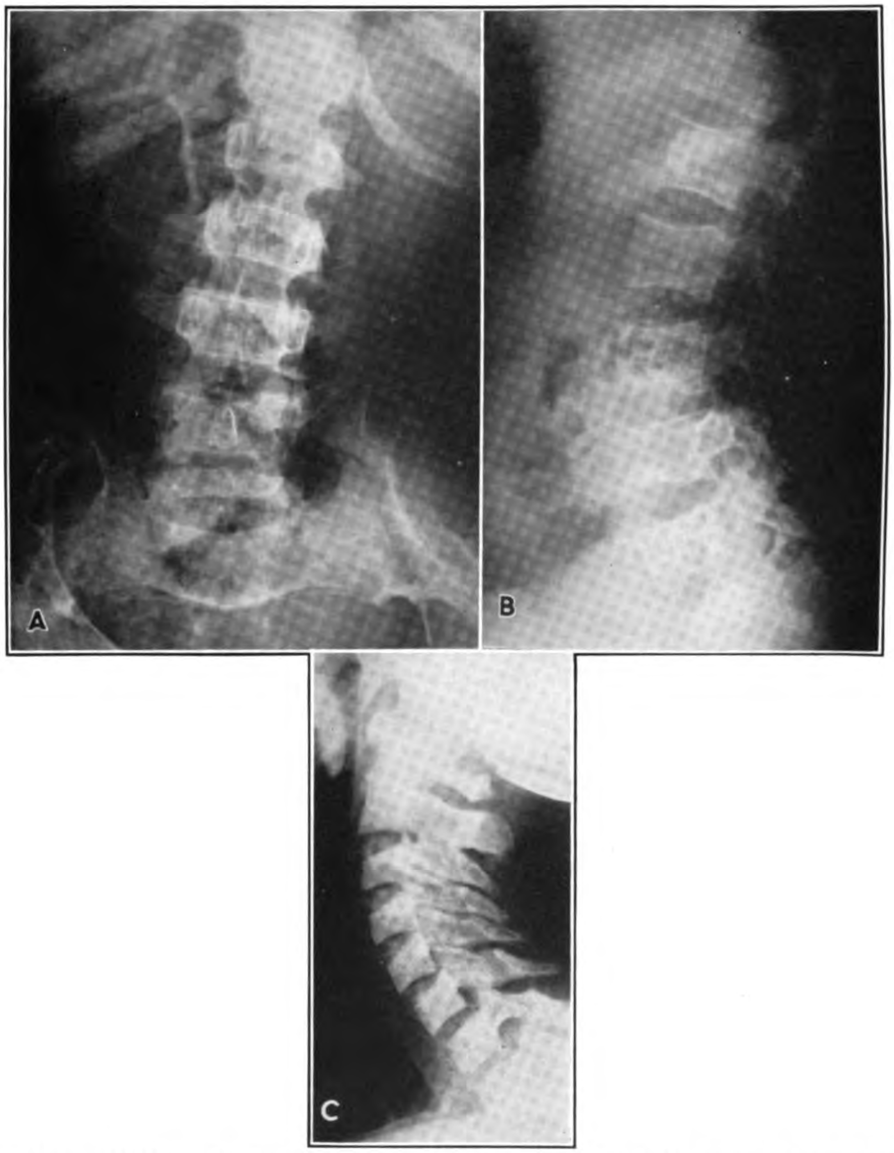

FIG. 108.—*A*, Fibrous dysplasia of bone in an eighteen-year-old male. Anteroposterior roentgenogram of lumbar spine. The vertebral bodies are moderately osteoporotic, and contain many fine, linear streaks of condensed bone. *B*, Same case, lateral view of lumbar spine. The intervertebral discs are moderately increased in height and are biconvex. Note the reticular appearance of the vertebral bodies. *C*, Same case. Lateral view of the cervical spine. The vertebral bodies appear denser than normal, and their trabecular structure is coarsened.

Other single instances were recorded by Warrick (1949) and by Schlumberger (1946).

I have had the opportunity of following a young man with this condition from the ages of 18 to 24 years. In addition to the classical changes in his extremities and the skin pigmentation, he had extensive involvement of his entire spine which remained relatively constant in appearance. The changes consisted mainly of a thinning of cortical bone, loss of trabecular markings and somewhat widened intervertebral discs. The cervical vertebral bodies were somewhat increased in density, with slight narrowing of their anterior aspects. A slight forward shift of the fifth on the sixth cervical vertebra was present, probably due to the increased lordotic cervical curve. The bodies of the thoracic and lumbar vertebræ had diminished density, and there were numerous vertical striations traversing the medullary spaces. The superior and inferior surfaces presented a biconcave appearance. The neural arches were somewhat demineralized but not deformed. In this patient the fifth lumbar vertebra was sacralized, sinking deeply into the superior surface of the sacrum. The transverse processes of the fourth lumbar vertebra were lower than the iliac crests and were directed upwards. This was probably due to progressive softening of the bone and the strains of standing and walking. At no time did this patient complain of spinal pain, nor was there any indication of central nervous system irritation.

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ARTHROGRYPOSIS MULTIPLEX CONGENITA

This term is used to designate multiple contractures of the joints, the term literally meaning crooked or bent joints. Other names by which this condition has been designated include amyoplasia congenita, congenital multiple or rigid joints, and arthromyodysplastic syndrome. The disease is of a hereditary familial nature and the characteristic changes are identified in newborn infants. We have had 2 instances in which the diagnosis was suggested on roentgenograms made before delivery in women with mild hydramnios by the peculiar stick-like extension of the legs on the femurs. Mild forms of the disease are consistent with longer, normal life.

The changes in the joints are primarily of a non-articular nature. The primary pathology in this condition is an atrophy and fatty replacement of the involved muscles with thickening of the periarticular structures and multiple contractures. An abnormality in anterior horn cells in the spinal cord and a denervation atrophy of muscle has been reported. The affected extremities assume an appearance which has been referred to as resembling a "stuffed sausage." As a rule, the involvement is symmetric, involving both the upper and lower extremities. Associated deformities such as myelomenenigocele hypoplasia of the jaw, congenital malformations of the hands and feet, disturbances of pigmentation of the skin and hair, congenital deafness have been reported.

The bony structures of the extremities characteristically present an elongated thinned-out appearance with adequate markings differentiating the medullary canal from the cortex. In 2 of our cases pathologic fractures occurred in the shafts of the bone, in 1 a concomitant of delivery. In another case, the femurs showed evidences of fractures *in utero* with healing, while the bones of the legs presented a slender appearance with a considerable degree of bowing secondary to softening of the bones. Bilateral clubfeet was observed in 3 of the 4 patients, together with congenital strictures of the wrist joints in 2 of these.

Involvement of the spine was considered as a secondary manifestation of the changes in the adjacent soft tissues. In a report of eighteen cases, Katzeff (1943) mentioned that in 4 cases the spine was affected. In the 4 cases observed by us, a lateral curvature of the spine was observed in 3 cases, of which 2 were newborns and 1 was a child one year and seven months of age. In another adult patient who presented all of the classical stigmata of arthrogryposis, including bilateral clubfeet and bilateral clubhands, together with an old congenital dislocation of one hip, the bony structures of the respective vertebræ were intact but again a curvature of the spine convex to one side was demonstrable. As mentioned by others, the changes in the alignment of the vertebral column represents in all probability a change in muscle pull rather than any disturbance of the bone itself.

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Osteopoikilosis

This condition is also known as osteopathia condensans disseminata, osteopoecilia, osteitis condensans disseminata, and spotted bones. It is a hereditary disease of mesenchymal disturbance and is an affliction characterized by the presence of multiple small round or ovoid foci of increased density scattered throughout the skeletal system, affecting chiefly the long bones and the pelvis. Involvement of the skull is quite rare. These small increased bony concentrations are very much like small osteomas and microscopically they are found to consist of densely packed bony tissue (Hinson, 1939).

In most of the cases reported the disease had been discovered incidentally and has not been accompanied by any symptoms referable to the islands of condensed bone. However, congenital anomalies associated with osteopoikilosis have been described, an example of which is a case of coarctation of the aorta associated with osteopoikilosis reported by Phalen and Ghormley (1943).

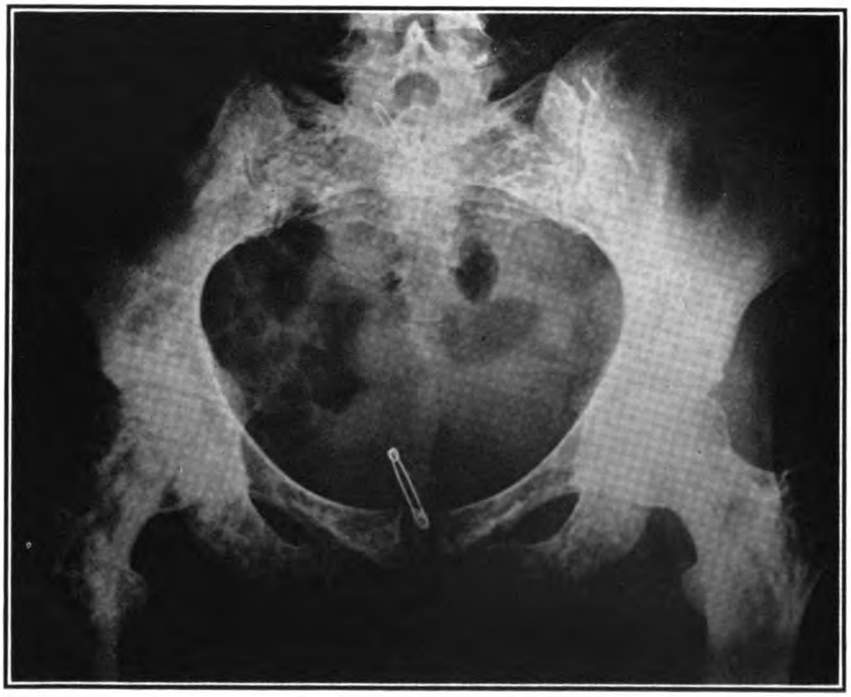

FIG. 109.—Osteopoikilosis in an adult female patient.

The spine is rarely involved in this condition and the published reports indicate only an occasional island in the lower lumbar spine or in the sacrum. In the case observed by us, a considerable degree of involvement of the sacrum was observed together with similar involvement in the pelvis, particularly in the pubic regions, and in both upper femurs. In addition, islands of compact bone were demonstrable in the fourth and fifth lumbar vertebræ. These were rather conspicuous in the transverse processes and neural arches.

The location of the multiple dense spots is within the cancellous bone. These appear as circular or ovoid shadows with their long axes usually parallel to the long axis of the affected bone. No such deposits have been observed in the cortex.

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OSTEOPATHIA STRIATA

This is an unusual affliction characterized by striation of the skeleton, particularly the metaphyses of the long bones. It is a disease of unknown origin and probably congenital in nature. It has been suggested that osteopathia striata is related to osteopoikilosis (Hurt, 1953) and dyschondroplasia (Voorhoeve, 1924), the three disorders being variants of the same fundamental error. However, Fairbank (1950) hesitated to accept this view. There are no definite symptoms referable to this dysplasia of bone, except that the patients sometimes complain of vague aches and pains. The outstanding radiologic change includes vertical striations in all of the bones except the skull and clavicles. The striation as a rule affects the shafts of the long bones, appearing as dense lines parallel to the long axis. The thickness of the striations may vary considerably and the bone in between various clusters of striated change may even be osteoporotic. The striation may or may not extend to adjacent epiphyses. The epiphyses are sometimes mottled with dense and clear spots. The spine is involved infrequently. In 1 of the cases reported by Fairbank, coarse vertical striations were present in the vertebral bodies.

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STIPPLED EPIPHYSES (Punctate epiphyseal dysplasia; chondroangiopathia calcarea seu punctata; chondrodystrophia calcificans congenita; chondrodystrophia punctata; chondrodystrophia calcarea congenita (Conradi's Disease).

This condition is a rare disorder of infancy of unknown origin, and may be familial. It occurs in about 1 in 500,000 births, and is perhaps more frequent in premature infants. The prognosis is poor, few surviving beyond two years. Nevertheless, instances have been recorded in which the patients reached adolescence or somewhat beyond this age. In some, no definite symptoms are observed, but more often associated changes such as shortened extremities, dry scaly skin, contractures of joints in flexion, syndactylism, congenital cataracts, microcephaly, oxycephaly, mental retardation, cleft palate or congenital cardiac defects have been present. Haymes and Wangner (1951) reported stippled epiphyses in an achondroplastic dwarf.

The disease is systemic, involving the cartilaginous portions of the epiphyses. The disturbances of calcification simulate those of chondrodystrophia fetalis, representing a congenital enchondral disturbance of ossification. As a result the long bones are shortened, with thick bowed shafts and wide irregular metaphyseal ends. The signs common to the disease include micromelia, rhizomelia, cataracts and stippled epiphyses. The nasal bridge is flat, but the skull itself is not affected.

The characteristic lesion is stippling of the various epiphyses, as if they were ossifying from multiple centers. The spots vary considerably in size and number. Anatomically there is disturbance of vascularization with spotty, mucoid degeneration of the epiphyses, as a result of which the hyalin cartilage becomes fragmented. Each fragment in turn may serve as a site for the formation of additional cartilage which ossifies and becomes connected by intervening cartilaginous bands. When the infant survives the formation of bone from cartilage slowly returns to normal and the islands of stippled density disappear. Bones which apparently were unaffected later in life may show malformations like those of chondrodystrophy. Licht and Jesiotr (1957) reported an atypical relatively symptomless manifestation of the disease in a 24-year-old man in whom the vertebræ were flattened and contained numerous areas of increased density due to spotty calcific deposits located principally in the anterior aspects of the bodies arranged in two symmetric lines opposite the pedicles.

The vertebræ participate in this generalized affection of enchondral calcification. As a result, stippling of the bodies appears, which follows the same tendency to vanish if the child survives. In some instances an atypical form of ossification of the vertebral bodies has been described, consisting of a coronal cleft of chondroid tissue on either side of which is stippling and disturbed ossification. Instances in which the vertebral bodies were deformed because of structural weakness also are known, occurring principally in youngsters who had survived well beyond one year. With extensive involvement this has been observed in infancy as well. The changes bear some resemblance to those of Scheuermann's disease. Disturbances in the ossification of the cervical vertebræ occasionally are followed by changes in the usual lordotic curve. Persistence of stippling into adolescence also occurs.

The coronal cleft type of deformity is attributed to the persistence of an anterior and posterior ossification center which failed to unite, presumably because of enchondral disturbance. Calcific flecks in the intervertebral discs result from occasional deposits of ossifying tissue in these structures during the period of growth. With survival, there is the same tendency to resumption of a normal growth pattern.

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DYSPLASIA EPIPHYSIALIS MULTIPLEX.

This is a rare, possibly familial developmental error of unknown origin, occurring approximately equally in either sex. It involves several or many epiphyses, principally those of the hips, shoulders, ankles and less often the hands and feet. As a rule the condition becomes evident clinically when difficulty in walking, pain and stiffness of the joints of the extremities brings the patient to his physician. Examination discloses that some patients have shortening of the limbs, shortened thick fingers and hands and other joint deformities as well. Usually the spine is normal, but in Jansen's case the epiphyseal disturbance also involved the growth centers of the vertebræ. Wedging and irregularities of the surfaces of the vertebræ in the thoracic region have been observed.

The radiologic changes are most evident in the growing epiphyses, where flattening, irregularity of ossification and distortion of the bone ends are most evident. When growth ceases the bones ossify in a more normal pattern, but deformities produced during growth remain even though the bones resume an approximately normal density. Arthritic changes are frequent in later life, particularly in the weight bearing joints.

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PROGRESSIVE DIAPHYSEAL DYSPLASIA

The disease, also known as Engelmann's disease and osteopathia hyperostotica (sclerotisans) multiplex infantalis is characterized by a progressive symmetrical thickening of the shafts of the long bones, associated with retarded growth and weakness. Particularly affected are the humeral and the femoral shafts and the bones of the forearms and legs. The vault and the base of the skull may show a diffuse thickening which sometimes reaches considerable proportions. Involvement of the vertebral column in this condition is rare, but thickening of the bones in the vicinity of the sacroiliac joint has been mentioned. A case with involvement of the vertebral bodies as well as the long bones, the skull, ribs, and scapulas is mentioned by Cocchi in an infant with a markedly sclerotic form of this condition.

The bony changes are constant throughout life and may even increase in extent as the patient grows older. While in its early stages the disease may be accompanied by difficulties in gait, together with some muscular atrophy and tenderness, the tendency is for these to recede and for the individuals later to be free from symptoms even in the presence of marked skeletal alterations. However, in infants in which there is a markedly sclerotic form of the disease anemia may appear and be uncontrollable.

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METAPHYSEAL DYSPLASIA.

This is a rare familial condition characterized by clubbing of the metaphyseal ends of the long bones, which assume a widened, somewhat tapered appearance with thinning of the cortex proximally and distally. The long bones may become bowed, and transverse lines of increased density are observed crossing the widened areas. The origin of this condition is unknown. The thinning renders them susceptible to fractures and stress deformities. In the cases reported by Feld, Switzer, Dexter and Langer incomplete pneumatization of the paranasal sinuses and thickening of the dorsum sellæ with a small sella turcica were observed. These authors also noted that the vertebral bodies in one of their cases presented some lengthening in the anteroposterior diameter. Komins described marked hyperostosis involving the facial bones and the base of the skull, prognothism and deficient dentition. The spine in his cases revealed some loss of the normal dorsal and lumbar curves.

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ARACHNODACTYLY (Marfan's Syndrome)

This condition is reputed to be an inherited mesodermal defect which some believe associated with eosinophilic hyperplasia and pituitary hypofunction. The disease is characterized by an abnormally slender and elongated appearance of the patient, with unusually thin fingers which gives rise to the comparison with spider claws. Concomitant congenital anomalies are frequently present, among which are included a high arched palate, small genitalia, and congenital cardiac malformations of which dissecting aneurysm of the aorta is a common variant. Migraine, epilepsy and retinitis pigmentosa are also frequently seen with arachnodactyly.

The skeletal system in these patients may present evidence of diffuse osteoporosis in addition to the abnormally slender appearance of the long bones. No specific changes are visible in the spine, but scoliotic changes secondary to relaxation of the paraspinal musculature is not infrequent. The osseous system presents certain similarities to eunuchoidism in which the extremities are long and slender and the epiphyses remain open for an abnormally long period.

Infantile Cortical Hyperostosis

In 1945 Caffey and Silverman reported on a syndrome which they designated as infantile cortical hyperostosis. The condition is of unknown etiology, and usually manifests itself in infancy by a thickening of the periosteum in the shafts of the long bones of the extremities, the short bones of the hand, the clavicles, and particularly the mandibles. Similar changes have been observed in the ribs. No involvement of the vertebral column has been reported.

In the cases observed by us no changes other than those reported in the literature were observed.

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Mongolism

This is a form of mental deficiency of obscure origin which is evident at or soon after birth. In a completely typical instance the patient presents the so-called mongoloid appearance with slanting eyes, narrow palpebral fissures and frequently an epicanthus. The children are usually small for their age, definitely mentally retarded and have poorly developed musculature so that the lack of coordination soon becomes evident. The protruding tongue and drooling from the mouth is usually seen in the more advanced cases. The condition usually terminates in death before adolescence, many mongols being seen as infants and relatively few surviving beyond adolescence into adult life.

Mongolism is frequently accompanied by congenital malformation of the heart. The hands and feet are usually short and thick, and the hands often present a shortened and inwardly curved fifth finger. A tendency towards a bifid formation of the hands and feet is present.

Skeletal maturation may be delayed in mongolism. The head may be brachycephalic and not infrequently microcephalic with hypoplasia of the nasal bones and upper maxillæ. While the inwardly curved fifth finger often observed with mongolism is considered characteristic of the disease, the same change may be observed occasionally in cretins and achondroplasts. Caffey has pointed out that the actabular angles of the hips are reduced and the ilia are broad.

Retardation of epiphyseal development may or may not occur with mongolism. Aside from the skull changes, no gross alteration in bony configuration has been described. The vertebral column shows no specific change in the bony structure of the vertebræ, but scoliosis secondary to muscular relaxation is not uncommon. In some of the mongols with large protuberant abdomens, a moderate dorsal convexity of the spine may be observed. In the cases observed by us, no specific vertebral changes could be made out.

Spitzer, Rabinowitch and Wybar (1961) noted that in 9 of their 29 mongoloid patients from 8 to 49 years of age that there was a forward displacement of the atlanto-occipital joint, together with abnormally thin and small atlas vertebræ.

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MELORHEOSTOSIS

This is a rare bony disturbance characterized by an unusual distribution of hyperostosis dispersed along the sides of bones of the extremities comparable to tallow dripping down a candle. It is usually confined to a single limb, with the hyperostosis selecting only one side of the bone. Limitation of the movements of the joints of the affected limb has been observed in about half the reported cases (Fairbank, 1948). Scleroderma, with fibrosis and thickening of the muscles and other soft tissues may be responsible for this stiffness. Involvement of the spine is rare. Carpender, Baker, Perry and Outland (1943) reported an eighteen-year-old female in whom the lateral aspects of the vertebral bodies extending from the seventh thoracic to the first lumbar vertebræ showed the striations characteristic of melorheostosis. Masserini (1944) also noted vertebral involvement with a dense, patchy well circumscribed type of deposit of compact bone in the vertebral bodies without distortion or symptoms.

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ENDOCRINE DISTURBANCES

PRIMARY HYPERPARATHYROIDISM

Albright and Reifenstein (1948) defined primary hyperparathyroidism as a condition in which more parathyroid hormone is manufactured than required. The parathyroid glands usually exist as four discrete structures, two beneath the inferior aspect of each lobe of the thyroid. The underlying pathologic change may be either a diffuse hypertrophy of all the parathyroid tissue or the development of single or multiple adenomas. A small number of malignant parathyroid tumors associated with hyperparathyroidism also have been reported (Gentile, Skinner and Ashburn, 1941; Meyer and Ragens, 1943; Stephenson 1950). Certain chemical findings in the blood are characteristic of hyperparathyroidism. These include increased serum calcium content, decreased inorganic phosphorous content of the blood, excessive excretion of calcium in the urine, increased excretion of inorganic phosphorous in the urine, and in those cases in which the skeletal system is extensively involved, an increased serum alkaline phosphatase content. Albright and Reifenstein believed that some parathyroid adenomas were connected with situations which tended to lower the serum calcium level, thereby stimulating parathyroid hyperplasia with the transformation of multiple circumscribed "germinative centers" into adenomas, with loss of control of these centers by normal stimuli. Association of parathyroid adenomas with pituitary and pancreatic islet cell tumors has been mentioned.

Hyperfunction of the parathyroid glands does not affect the skeleton primarily, but first influences the phosphate metabolism by lowering the renal threshhold for the excretion of phosphorus in the urine. This in turn is accompanied by a fall in the blood serum phosphate. The serum calcium increases in order to keep the relationship between the serum calcium and phosphorus constant, thereby mobilizing calcium phosphate from the bone with consequent hypercalciuria. In addition to this mechanism Albright also believed that an excess of parathyroid hormone had a direct influence upon bone structures. Parathyroid hyperplasia and adenomas may occur without renal or osseous manifestations (Rogers, 1946).

The original description of osteitis fibrosa was made by von Recklinghausen, who considered the change a primary disease of the skeleton. The association of osteitis fibrosa with a parathyroid tumor was noted in 1904 by Askanazy (Snapper, 1949) and experimental evidence of the influence of the parathyroid glands on calcium metabolism was reported by Erdheim (1907). Erdheim believed that the hyperplasia of these glands was a compensatory mechanism to permit calcium mobilization from the skeleton replacing calcium lost in the urine and stool. In 1926 Mandl reported the first case successfully operated upon for the removal of a parathyroid tumor. Mandl had transplanted four parathyroid glands from a moribund patient to a patient who preoperatively showed extensive osteitis fibrosa. This was done to test Erdheim's concept that parathyroid hyperplasia was a compensatory mechanism. Following the operation the patient became worse, whereupon Mandl not only removed the transplanted glands but explored the patient's mediastinum and removed the offending parathyroid tumor. Operation was followed by a rapid cessation of clinical symptoms and return of the blood calcium to a normal level. The first case in this country of hyperparathyroid tumor with surgical removal of a lesion from the mediastinum was described by Hannon, Shorr, McClellan and DuBois (1930). The monograph by Shelling summarized all available data up to 1935.

In hyperplasia of the parathyroids the growth is chiefly of the clear cells. With adenomas the chief or principal cells multiply in acinar formations, with intervening areas of degeneration and small cyst formations. The presence of a small tumor of clear cells surrounded by a shell of normal parathyroid tissue is considered quite characteristic of an adenoma. The pathologic changes were described in detail by Castleman and Mallory (1935).

The clinical picture of hyperparathyroidism apparently is the same no matter whether the clear cells or the chief cells predominate. It is currently considered that the water clear cells of the parathyroid glands represent the stage of greatest hormonal activity. The chief, or principal cells, indicate a resting stage. Young chief cells, which are water clear transitional chief cells, are regarded as possibly being activated principal cells. The eosinophilic cells found in the parathyroid glands have no known endocrine activity. In a review of 63 cases, Black (1948) encountered single adenomas in 56 cases, multiple adenomas in 3, and diffuse primary hyperplasia in 4. Generalized osteitis fibrosa cystica was present in less than 40 per cent of his cases. Thirty-five per cent had renal complications, but no evident bone disease, and 20 per cent had bone changes without renal complications. About 20 per cent had both renal and bone disturbances. He considered the urinary changes more frequent and more important than the bony complications.

Hyperparathyroidism occurs in either sex, but is rather more frequent in females of middle age. However, cases have been observed in children and elderly individuals. The condition may exist in a subclinical form for several years, becoming evident only when manifestations secondary to demineralization of bone results in weakening of these structures, with consequent deformities and pathologic fractures. Coburn (1944) reported such a case followed for fifteen years. The patients may also develop a peculiar muscular atonicity due to hypercalcemia. Anorexia, nausea, vomiting and abdominal pain may also be noted. With the maintenance of hypercalciuria the urine may be supersaturated with calcium phosphate and stones appear. These sometimes become quite large and may be responsible for impaired renal function. Another type of deposition of calcium is so-called metastatic calcification, in which lime salt is laid down in various organs, especially the kidneys, and less often the heart, the lungs, and sometimes the corpora cavernosa and the mucosa of the gastrointestinal tract. When the kidneys are considerably infiltrated with such calcification the lime salt deposits can be identified on roentgenograms as nephrocalcinosis. The difficulties in establishing a diagnosis of hyperparathyroidism on biopsy material alone was stressed by Black and Ackerman (1950), who believed that the characteristic roentgenographic and chemical changes were also required to make the diagnosis. Secondary hyperparathyroidism due to renal disease may so simulate primary hyperparathyroidism that the two are indistinguishable (Curtis and Filler, 1942.)

The clinical picture of hyperparathyroidism may be one which has to be differentiated from others which in part produce similar details. For instance, nephrocalcinosis may be seen with chronic pyelonephritis or glomerulonephritis, which in turn often results in the indistinguishable secondary form of hyperparathyroidism. Other causes for nephrosclerosis which are more readily distinguished include vitamin D intoxication, hyperchloremic acidosis, sarcoidosis, idiopathic hypercalcuria, excessive osteolytic activity such as may be associated with multiple myeloma or extensive bone metastases, bone tuberculosis or the milk-alkali syndrome observed in patients with peptic ulcers undergoing rigorous treatment. The association and persistence of peptic ulcers in patients with hyperparathyroidism alone or as part of multiple endocrine adenomas is worthy of special note, since some of the resistant patients may have hyperparathyroidism without the more readily detectable features. Hypertension with hyperparathyroidism is related to the underlying renal damage.

The skeletal changes in hyperparathyroidism are caused by osteoclastic activity which produces rapid decalcification, leading to osteitis fibrosa, a nonspecific reaction to rapid demineralization. Cyst formation, and

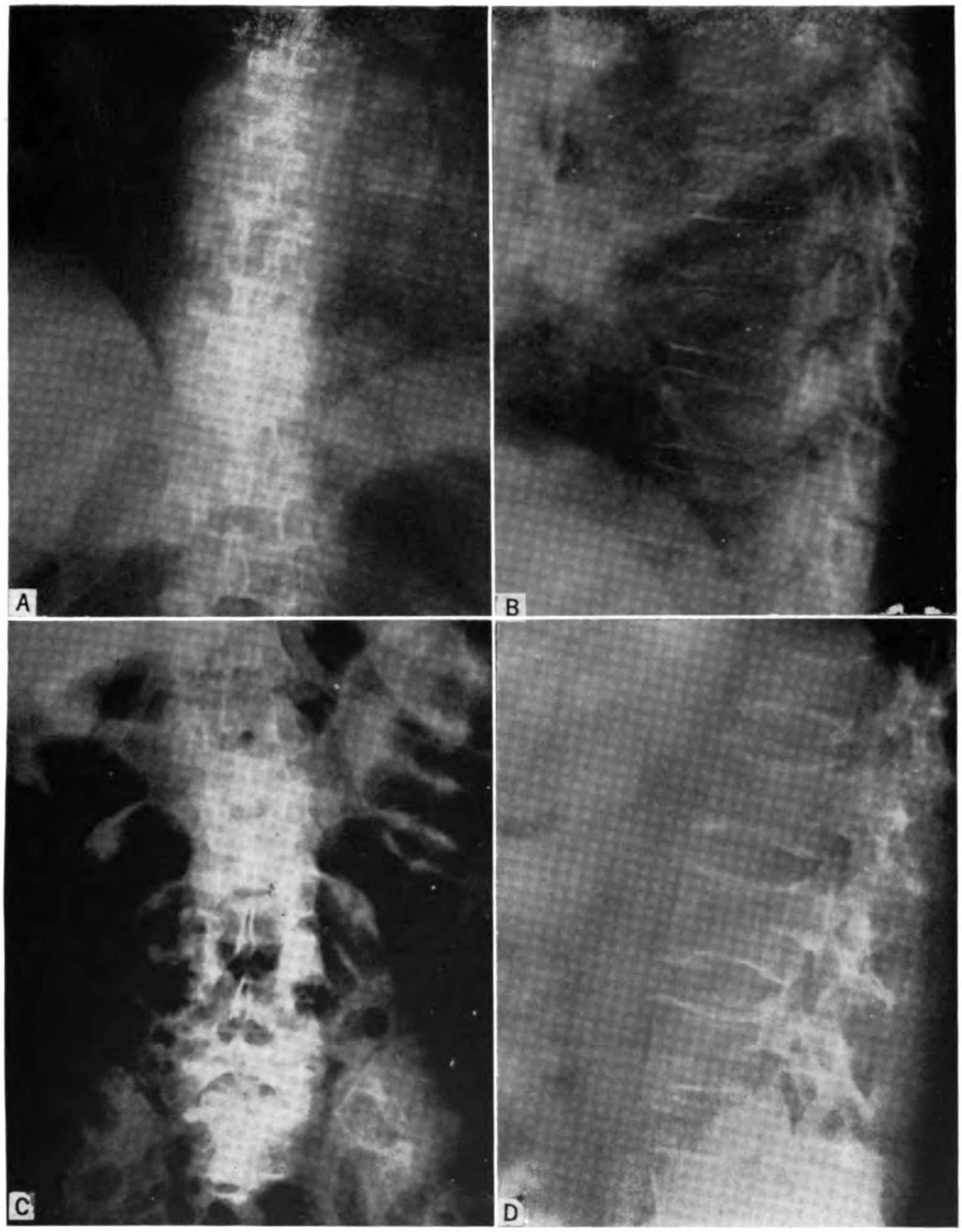

FIG. 110.—Spinal roentgenograms of a 48 year old woman with hyperparathyroidism, verified at operation. *A* and *B*, thoracic spine as well as the lumbar spine *C* and *D*, reveal pronounced demineralization of bone. The vertebral bodies in the thoracic area are somewhat flattened, especially those in the midthoracic region. The lumbar intervertebral discs are bulging, presenting a biconvex appearance.

brown giant cell tumors, the so-called osteoclastomas, are found. The increased parathormone causes proliferation of osteoclasts in the bone marrow with erosion of trabeculæ followed by decalcification and osteitis fibrosa. The rise in blood and urinary calcium is a result of the bony disturbance. Because of the increased phosphate excretion by the kidneys, hypophosphatemia develops. The rise in phosphate renal excretion is due to excess parathyroid hormone. Whether this hyperphosphaturia is simply due to increased glomerular filtration or to decreased phosphate resorption in the renal tubules, or to a combination of both conditions is still undecided. No agreement has been reached as to whether a decreased phosphate reabsorption is due to a direct parathyroid hormone influence on the tubules, or whether hypercalcemia irrespective of its cause can decrease tubular reabsorption of phosphates. There is reason to believe that the demineralization in hyperparathyroidism may depend on still unknown mechanisms (Snapper, 1953).

From a roentgenologic viewpoint, hyperparathyroidism is accompanied by a diffuse form of osteoporosis which may involve the entire skeleton. In those areas in which bone resorption is most pronounced the cortex of the bone may be thinned out, presenting a scalloped appearance. Adjacent thereto the bones may be expanded and there may be numerous connecting areas of radiolucency presenting at times an almost web-like appearance. Considerable deformity of the long bones secondary to softening and pathologic fractures is not infrequent. The skull may show a miliary form of decalcification and occasionally this assumes a fairly sharply delimited appearance resembling osteitis circumscripta. The demineralization of the skeletal structures may proceed fairly rapidly, and in the most advanced forms there are large cyst-like regions of demineralization criss-crossed by residual bands of fibrous tissue. In other instances of hyperparathyroidism the cortex of the bone presents numerous small sharply circumscribed areas of decalcification, which later become confluent with ill-defined borders. The lamina dura around the teeth disappear. Another helpful sign is the identification of ungual tuft resorption, and more often subperiosteal decalcification with small vertical spicules of bone produces a so-called palisade formation (Pugh, 1952). This also occurs with secondary hyperparathyroidism, as does trabecular resorption of spongy bone at areas of stress such as the symphysis pubis and the distal ends of the clavicles.

In several cases of hyperparathyroidism which we have observed in adults who had been bedridden for several years, in addition to the cystic changes in the bones there was marked sharpening and pointing of the distal ends of the radius and ulna and of the small bones of the hands, with marked dislocations of these structures because of the complete loss of articular structures.

Roentgenologic manifestations of osteitis fibrosa cystica in the spine may be divided into two varieties. The more common form is the one in which the osteoporosis is pronounced, with demineralization of the entire vertebral column resulting in the formation of a dorsolumbar or dorsal kyphosis with shortening of the height of the patient and a forward protrusion of his head and neck. The vertebral bodies may collapse in a uniform fashion so that the normal height of the affected vertebral body is reduced, leaving the articular surfaces relatively smooth and the intervertebral spaces about average. This occurs usually in the dorsal spine. In the lumbar spine the osteoporosis may be manifested by a progressively more marked biconcave appearance of the vertebral bodies, and a consequent biconvex increase in the height of the intervertebral discs. With softening the protrusion of nucleus pulposus material into the vertebral bodies may become quite prominent. In such cases there is rarely any bony reaction around the herniated nuclear material. While neither change described above is characteristic of hyperparathyroidism, that possibility must be kept in mind and further evidence sought both clinically and in radiographic studies of other skeletal structures and the kidneys.

A second and less frequent form of spine involvement with hyperparathyroidism is that of a reticular web-like change with relatively sharply defined lacunæ together with cyst-like lesions of varying sizes and configurations. In 1 of the cases observed by us this was particularly prominent in the cervicodorsal region, with similar alterations in the adjacent ribs. The long bones in this patient were of normal bony density but contained numerous small and moderately large sharply circumscribed cysts. There was a diffuse collapse of the body of the seventh cervical vertebra. Interspersed in the other osteoporotic cervical segments were numerous small fairly sharply defined islands of dense bone. In this patient it was also of interest that the softening of the ribs had apparently lowered the thoracic inlet so that the first and second dorsal vertebræ were clearly outlined above the first rib shadows.

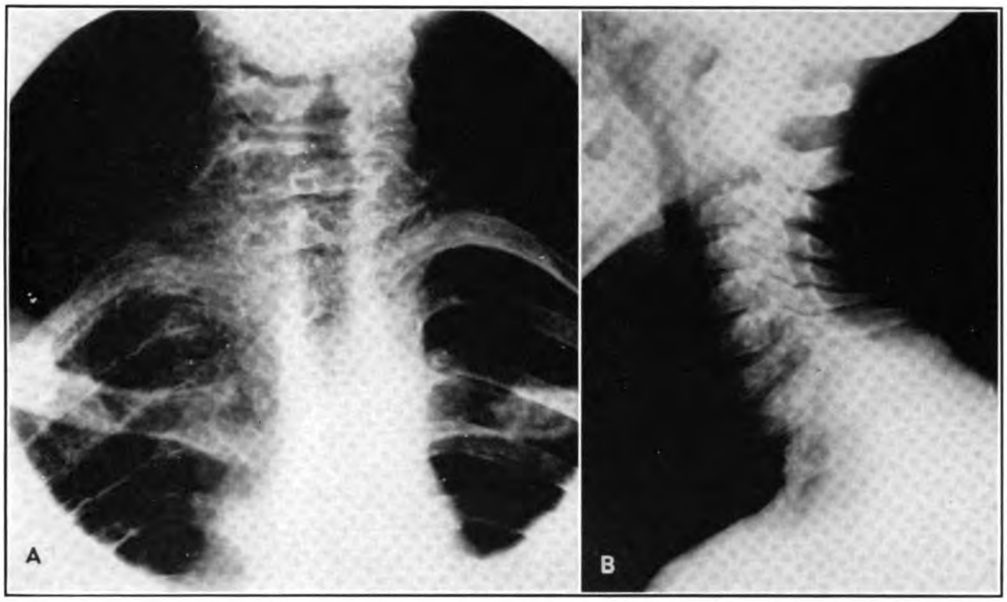

FIG. 111.—Hyperparathyroidism. The upper right ribs and the vertebræ are involved in a reticular osteoporotic pattern of osseous change, which extends to the neural arches as well.

Progressive osteitis fibrosa cystica may cause decalcification of the sacrum and practically complete obliteration of the sacroiliac joints. The width of the sacrum may become narrowed in its transverse diameter as a consequence of pressure of both iliac bones.

While the usual effect of either primary or secondary hyperparathyroidism is one of loss of bone, instances in which osteosclerosis of the vertebral bodies occurred with this condition are being reported with increasing frequency. The cause for this bizarre change is unknown.

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Secondary Hyperparathyroidism

Under secondary hyperparathyroidism are included conditions which result in parathyroid hyperplasia with formation of excess parathyroid hormone. It may occur with or without the characteristic chemical or bony changes, and with histologically normal glands (Soffer and Cohen, 1943). Chief among these conditions is chronic renal disease, leading to the designation of the disorder as renal osteodystrophy. Such renal disease may follow primary urinary tract disturbances such as chronic glomerular nephritis, pyelonephritis and nephrosis. Congenital malformations of the kidneys such as polycystic kidneys and renal hypogenesis may also be causative. Other urinary tract disturbances such as obstructions due to urethral valves, prostatic hypertrophy, urethral strictures and bladder neck or ureteral strictures may also be responsible for such changes. A rare cause of secondary hyperparathyroidism may be the primary renal tubular disorder called Fanconi's syndrome, in which there is acidosis, hyperchloremia, glycosuria and an abnormal excretion of organic acids in the urine with a low serum phosphorus content. Another form of tubular damage may result in hyperchloremia without glycosuria (Rule and Grollman, 1944). Secondary hyperparathyroidism also may be due to calcium deprivation due to diet, pregnancy, lactation, rickets and osteomalacia. Idiopathic hypercalcuria is another form of so-called vitamin D resistant rickets, with increased calcium excretion of unknown etiology.

Renal insufficiency may be accompanied by abnormal serum phosphate and nonprotein nitrogen retention, and normal or slightly diminished serum calcium content. A high serum phosphatase level is present, as well as acidosis. Hyperplasia of the parathyroids follows the low serum calcium content which compensates for the elevated phosphate in the blood. The parathyroids in such patients, according to Castleman and Mallory (1935), may either remain average in size or increase markedly in bulk. There is a prominent increase in the number of normal-sized chief cells, with no mitotic activity. A diminution or absence of intercellular fatty tissue appears, and there is a higher glycogen content than may appear with parathyroid hypertrophy or adenomas. Such a hyperplastic gland, as occurs with secondary hyperparathyroidism, may be greatly increased in its parathyroid hormone productive capacity because of the prominence of glandular tissue replacing the normal intercellular fat.

Secondary osteonephropathy may exhibit pronounced bone changes, particularly in children. These individuals may be markedly deficient in growth, resulting in the picture of renal dwarfism or renal nanism. The epiphyses may present a marked degree of widening, with irregularities of their margins which resemble advanced rickets, leading to the designation of this condition as "renal rickets."

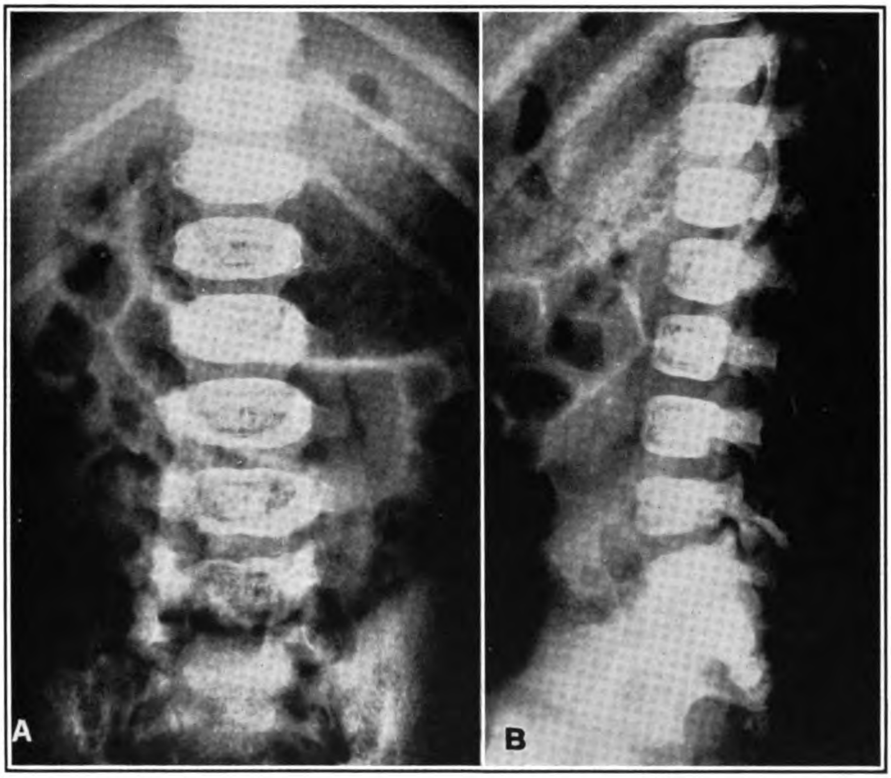

FIG. 112.—Renal osteodystrophy due to congenital obstruction at the vesicle neck of a nineteen-month-old boy. The vertebral bodies present a dense core around which is a layer of radiolucent structure, which in turn is enveloped in a thin dense periphery.

It has been suggested by Albright and his co-workers and by Snapper that this term be discarded. Albright pointed out that the epiphyseal lesions of renal rickets are quite unlike those seen with true rickets, and that hyperparathyroidism with osteitis fibrosa generalisata uncomplicated by kidney damage is not associated with epiphyseal lesions in children. A pronounced degree of osteoporosis of the entire skeletal system may be observed, with a translucency of the bones of a much more pronounced type than is usually observed with rickets. Subperiosteal elevation may occur in the ends of the long bones and in the hands and feet. The epiphyseal discs in these patients may remain open for an unusually long time even if the patient reaches adolescence or adult age. The bone changes with renal disease were reviewed by Cancelmo and Bromer (1951).

In the case illustrated here (Fig. 112) the patient was a nineteen-month-old boy with congenital obstruction at the vesical neck. This had remained untreated for more than a year because his parents refused to heed medical advice. The child was admitted with acute urinary retention, with a urea nitrogen content of the blood varying between 95 and 100 milligrams per cent, a blood calcium of 10, alkaline phosphatase of 6 and an inorganic phosphorus content of 6.1 milligrams per cent. This patient presented a classical picture of renal osteodystrophy, with subperiosteal absorption at the distal ends of the humerus and forearms, an irregular appearance of the resorptive bone at the distal ends of the clavicles and generalized stunted growth. The lamina dura of his teeth were deficient, but the skull itself showed no gross change.

Roentgenographic examination of his spine was of particular interest. This revealed the spinal column to be normally aligned. The vertebræ were of normal height, as were the intervertebral discs. The vertebral bodies maintained their normal proportions with a tendency towards slight rounding of the articular margins of the upper lumbar vertebræ. No wedging was visible. The vertebral bodies presented a peculiar reticular decalcification immediately beneath their superior and inferior surfaces. To some degree this change was present along the anterior and posterior aspects of the vertebral bodies. We had available for comparison roentgenograms when the patient was six months of age at which time the skeletal changes were minimal, being limited to the epiphyses, and the vertebral bodies appeared to be within average limits. Apparently the time necessary for pronounced bony changes to develop in this patient had been approximately a year or less.

Renal osteodystrophies in adults do not present as prominent changes in the skeletal structures. Visible changes do not occur at the ends of the long bones because the epiphyses are united. The condition may manifest itself more in the nature of metastatic calcinosis, with deposits in the soft tissues, and Mönckeberg's form of medial arteriosclerosis, together with a varying degree of osteoporosis which sometimes may be extreme. Such a case was reported by Dreskin and Fox (1950) in a twenty-seven-year-old man with chronic renal disease since childhood, osteoporosis, parathyroid hyperplasia and metastatic calcification. The unique feature in this case was the predominant enlargement of one parathyroid gland simulating a palpable adenoma. The pathologic report included a description of tissue from the bones, including the vertebræ. These showed islands of hematopoiesis, but the major portion of the marrow space was occupied by large numbers of irregular trabeculæ of cross-fibrillar bone lying in a fibrous matrix. The spongiosa showed predominantly osteoblastic new bone formation with rows of plump osteoblasts aligned along the trabeculæ, and the spicules had moderately wide osteoid margins. There was no apparent delay in calcium salt deposition in the osteoid material. Osteoclastic activity was also seen, with multinucleated osteoclasts lying in wide lacunar spaces. This lytic process was accompanied by deposition of new bone. The cortices were thin throughout, with enlarged arlacun spaces containing fibrous tissue. Here, too, new bone was being laid down. The fibrous tissue was vascular and moderately dense, and there was no evidence of inflammatory reaction in this case.

Renal osteodystrophy secondary to congenital polycystic hypoplasia of the kidneys associated with diabetes and polycystic disease of the pancreas was described by Jackson, Bates, Slavin, and McFarland (1950). The patient was a twenty-five-year-old man whom these authors considered to be unusual because of the presence of diabetes and because the blood nonprotein nitrogen and serum phosphorous levels were elevated. In this patient the lumbar spine showed moderate osteoporosis as did the skull. The long bones showed cortical thickening with the appearance of increased trabeculations and multiple tiny cysts. In a study of the vertebræ of 39 patients who had died of renal insufficiency, Follis and Jackson (1943) found histologic changes in about half. The principal abnormality was excessive osteoid tissue rather than osteitis fibrosa.

A case of adult renal osteodystrophy was included in the report of Albright, Drake and Sulkowitch (1937) in which a post-mortem roentgenogram of the spinal column revealed decalcification and herniations of the nuclei pulposæ into the vertebræ.

A rare form of osteonephropathy is cystinosis, the Lignac-Fanconi syndrome. In this condition one encounters glycosuria without hyperglycemia, acidosis, a low serum phosphorus content and an abnormal excretion of organic acids in the urine. These patients present a picture of dwarfism and rickets which is considered to be due to a congenital malfunction of the kidney tubules, so that for some unknown reason there is a failure of reabsorption of glucose, amino acids and phosphates. As a result these substances are excreted in abnormal amounts, producing amino-aciduria with normal amino acid blood levels, glycosuria with normal blood sugar level and excess excretion of phosphorus in the urine. An excessive unidentified acid is present in the blood but an increased production of ammonia by the kidneys keeps the urine alkaline. The osteomalacia produced in this condition appears to be the result of excessive loss of phosphorus in the urine. Radiologically it is difficult to distinguish this condition from advanced infantile rickets. Sub-periosteal resorption of bones in the hands does not occur. Osteomalacia of the vertebral column has been described in association with generalized osteoporosis.

The problem of rickets and osteomalacia from renal tubule defects was reviewed recently by Dent (1953) who suggested the designations of glomerular and tubular rickets, recognizing that some cases of tubular rickets terminated in uremia and were indistinguishable from primary glomerular rickets. Tubular damage, by the same token, was present in cases of glomerular rickets. Recent biochemical advances in his opinion justified attempts made by him to reclassify the forms of tubular rickets, namely, those forms resistant to vitamin D therapy in which the renal dysfunction was mainly concerned with the various known actions of the renal tubules. He stated that there was an adult as well as a childhood form of renal tubular defect with similar biochemical disturbances, and that the differential diagnosis of the various tubular disturbances was largely a chemical matter. The roentgenologic changes in the various forms of tubular disturbances were identical, and might occur at any age, in either sex, and in acute or chronic form.

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HYPOPARATHYROIDISM

This condition is due to a deficiency in the production of parathyroid hormone. The most common cause is accidental injury to the parathyroid glands, or accidental removal in the course of thyroid surgery. Rare cases of idopathic hypoparathyroidism occurring without known reason have been described. This condition is associated with tetany, characterized by muscular cramps, carpopedal spasm, convulsions and laryngeal stridor. Cataracts are also frequently observed with hypoparathyroidism. A frequently observed manifestation of hypoparathyroidism is the presence of calcification in the basal ganglia of the brain, which Albright and Reifenstein regard as evidence that the change in hypoparathyroidism are in the direction of supersaturation of body fluids with calcium phosphate. However, such basal ganglia calcifications are not necessarily pathognomonic of hypoparathyroidism. Hypoparathyroidism is associated with hypocalcemia and hypophosphatemia.

In so far as I can determine there are no changes in the spinal column which may be attributed specifically to this condition.

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PSEUDOHYPOPARATHYROIDISM (Seabright Bantam Syndrome)

This condition is differentiated from true hypoparathyroidism by the fact that in pseudohypoparathyroidism the disturbance is an inability to respond to the presence of normal parathyroid hormone. This syndrome was first described by Albright, Burnett, Smith and Parson (1942), who reported that these patients had a tendency to be unusually thick-set and short with a tendency toward brachydactyly, especially in the metatarsal bones, because of early closure of the epiphyses. Considerable soft tissue calcific deposits may be observed, some of which can be felt on physical examination. Tetany is present, with low serum calcium and elevated serum inorganic phosphatase. The term "Seabright Bantam Syndrome" was suggested by a comparison to this breed of chickens in which the roosters present a feminine type of plumage, presumably because of an insufficient response to male sex hormone indicating, the presence of an end-organ refractory state.

In the cases reported the long bones reflect the dwarfism present. Calcifications in the basal ganglia as well as elsewhere in the soft tissues have been described. The skull bones are thickened. The bony changes in the long bones sometimes are like those of an osteochondrodystrophy, with marked epiphyseal irregularities and softening and malformations which tend to return to a normal state when growth ceases. Changes in the vertebral column are not striking, consisting mainly of some demineralization of bone with accentutation of the vertical trabeculæ, some irregular calcification within the bodies and interspersed dense and rarefied areas.

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PSEUDO-PSEUDOHYPOPARATHYROIDISM.

There exists a small group of patients with some of the changes seen with hypoparathyroidism, but who have neither hypocalcemia nor hypophosphatemia, and consequently no tetany or convulsive symptoms. However, these patients present changes of dyschondroplasia in their short bones of hands and feet, round faces, short stature and metastatic calcifications in the soft tisues. No basal ganglia calcification has been encountered in this disorder.

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CUSHING'S SYNDROME

Cushing's syndrome is characterized by increased trunk fat, purplish stripes on the abdomen, a full moon face, hirsutism and acneiform eruptions, hypertension and decreased sugar tolerance. The skin becomes thin and dry, with cyanosis of the face, hands, neck and feet. The patient may be irritable and drowsy, with marked muscular weakness. Polydipsia, polyphagia and polyuria may be noted. A polycythemia of mild degree may be present. A low basal metabolic rate is noted in many of these patients, but this may be normal or even somewhat elevated. Arteriosclerosis and nephrolithiasis are common concomitants of the disease. In females amenorrhea is frequent, and in the male loss of libido and impotence is observed. This clinical picture was originally developed by Cushing (1932), who ascribed its cause to basophilic adenomas of the pituitary gland. Eisenhardt and Thompson (1939) called attention to the frequency with which osteoporosis was present with pituitary basophilism. Later, Cushing's syndrome was found to be due to other causes including various adrenal cortical tumors, tumors of the ovary, tumors of the thymus or atrophy or inhibition of the hypothalamic nuclei, particularly of the paraventricular and the supraoptic nuclei. Heinbecker (1946) reviewed experimental evidence which indicated how these four primary causes depressed the function of the neural hypophysis and affected a dominance of the eosinophilic adrenal cortical hormone complex over the basophile thyroid complex.

The diabetes associated with Cushing's syndrome is usually resistant to insulin. Atrophy of lymphoid tissue with lymphocytopenia and polymorphonuclear leucocytosis is also present. When the condition occurs in children retardation of skeletal growth takes place. The urinary excretion of 17-hydroxycorticoids usually is elevated.

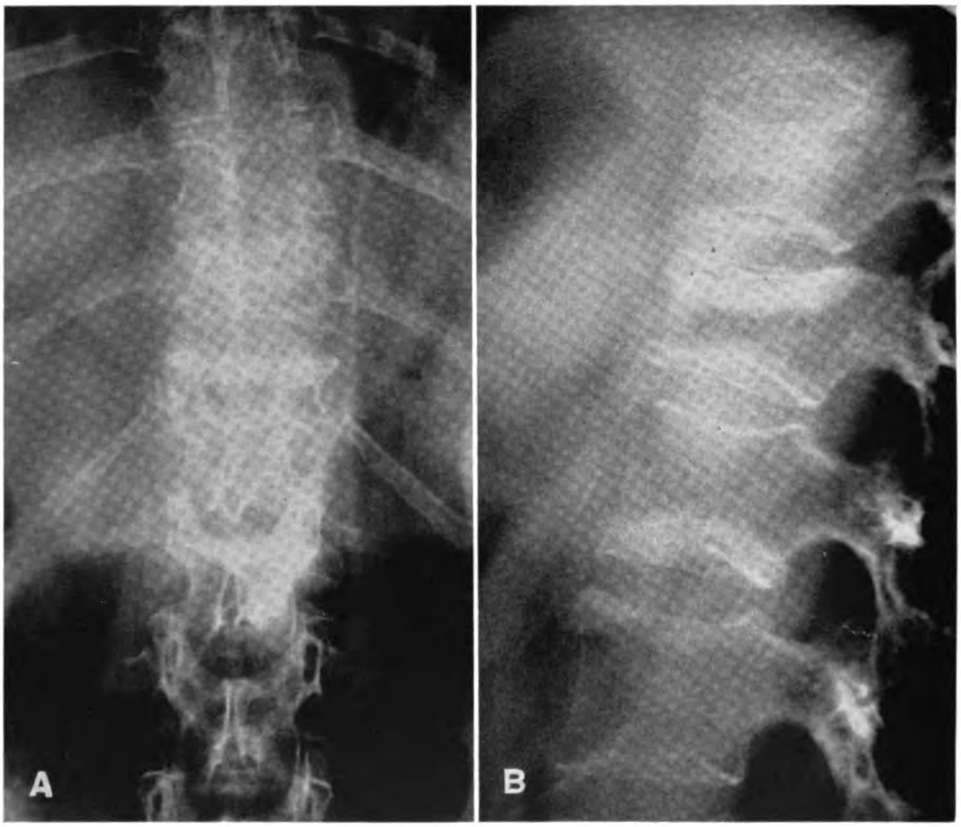

FIG. 113.—Cushing's syndrome in a 46-year-old-woman. The vertebræ are osteoporotic, and compressive changes with sclerotic margins are present in the superior surfaces of the twelfth thoracic and first lumbar vertebra. Similar changes are seen at the inferior aspect of the second lumbar vertebra. The intervertebral discs are bulging.

According to Albright and his co-workers the immediate cause of Cushing's syndrome is an overproduction of the adrenal cortical "sugar" or "S" hormone. In many cases the overproduction of this hormone is due to cancer of the adrenal gland. In those without cancer of the adrenal the cause of the overproduction is not clear, and Albright believed that it may be due to excess amounts of adrenocorticotropic hormone. Albright and his co-workers further discussed the role of the so-called nitrogen or "N" hormone, which they regarded as opposite in its action to the sugar hormone. They believed that an excess of nitrogen hormone should neutralize an excess of sugar hormone, which to a great degree forms the basis for treatment of osteoporosis with testosterone, whose action is very much like that of the "N" hormone.

Knowlton (1953) questioned whether the pituitary, the adrenal cortex or the hypothalamus was the site of the original lesion, although it was generally agreed that the end result is due to excess activity of the adrenal cortical hormones.

Similar conditions appear in patients treated over long periods with ACTH or cortisone. In a review of 189 cases from the literature and 5 cases of his own, Knowlton found that the clinical picture was variable, that the hypertension was reversible and that the suppression of gonadal function was not universal. He found *x*-ray evidence of spinal demineralization in 74 per cent of cases, with compression fractures in 34 per cent. Skull osteoporosis was present in 50 per cent and rib fractures in 11 per cent.

From the radiologic viewpoint the osseous manifestations of Cushing's syndrome are principally those of osteoporosis, due to disturbed protein metabolism leading to a lack of bony matrix formation and deficient osteoblastic activity. The spine is effected, to a major degree.

Cushing's syndrome is associated with widespread osteoporosis which affects the spine, the ribs, pelvis and skull most severely. The vertebræ become demineralized, particularly so when the disease has been present for a long time. The peripheral cortical bone forming the walls of the vertebral bodies is thinned, but because of the loss of cancellous bone the margins appear relatively dense. Increase in radiolucency of affected vertebræ results from trabecular loss. The vertical trabeculæ appear to persist better than the horizontal ones, so that not infrequently the vertebræ have relatively prominent vertical markings. Further changes are those incident to strain and weight bearing, and appear most often in the thoracic and lumbar regions. The affected vertebræ may become biconvex, with bulging intervertebral discs producing exaggerated saucer-like indentations into the upper and lower surfaces of the bodies. Sometimes a sclerotic reaction appears along the margins of the bone in contact with the discs. Nuclear intrusions into the vertebræ may be quite deep, and can produce faintly sclerotic reactions in the adjacent cancellous bone. Further loss of structure strength results in bulging of the vertical walls of the vertebræ or wedging or flattening of the bodies. In some the deformity is mild, while in others the malformation is so severe that the vertebræ are almost discoid or deeply biconcave in configuration.

The pathological appearance of vertebræ affected by Cushing's disease was described by Follis (1951) and by Sissons (1956). The principal change of osteoporosis is reflected by the diminished osteoblastic activity resulting in thin, delicate trabeculæ which may be angulated and sometimes crenated. Fatty replacement of hemopoietic tissue occurred in areas where trabeculæ were absent and where bony rarefaction and collapse were greatest.

As might be anticipated because of the effect of abnormal corticosteroid function, the amount of osteoid present about the trabeculæ is quite small. The increased amounts of adrenal hormone inhibits protein synthesis, thereby diminishing the production of bone from osteoid. Follis noted rather thin osteoid borders about many of the thinned trabeculæ in his patients. Sissons found that the surfaces of the slender trabeculæ were complete devoid of osteoid borders. Normal osteoblasts were seen, but these were reduced in number and their bone-forming activity appeared minimal. Osteoclasts were rare.

Sissons found numerous fractures of trabeculæ in those vertebræ which were collapsed. He further noted that in most areas the reaction of adjacent tissues was minimal, although in one vertebra the displaced trabeculæ in the collapsed area were surrounded by vascular granulation tissue and proliferating osteoblasts. Even though there was little reaction to the fractured trabeculæ, they did later join into irregular masses of bone. "The structural changes in these vertebræ (see for instance Figures 2, 4, 5 and 6)

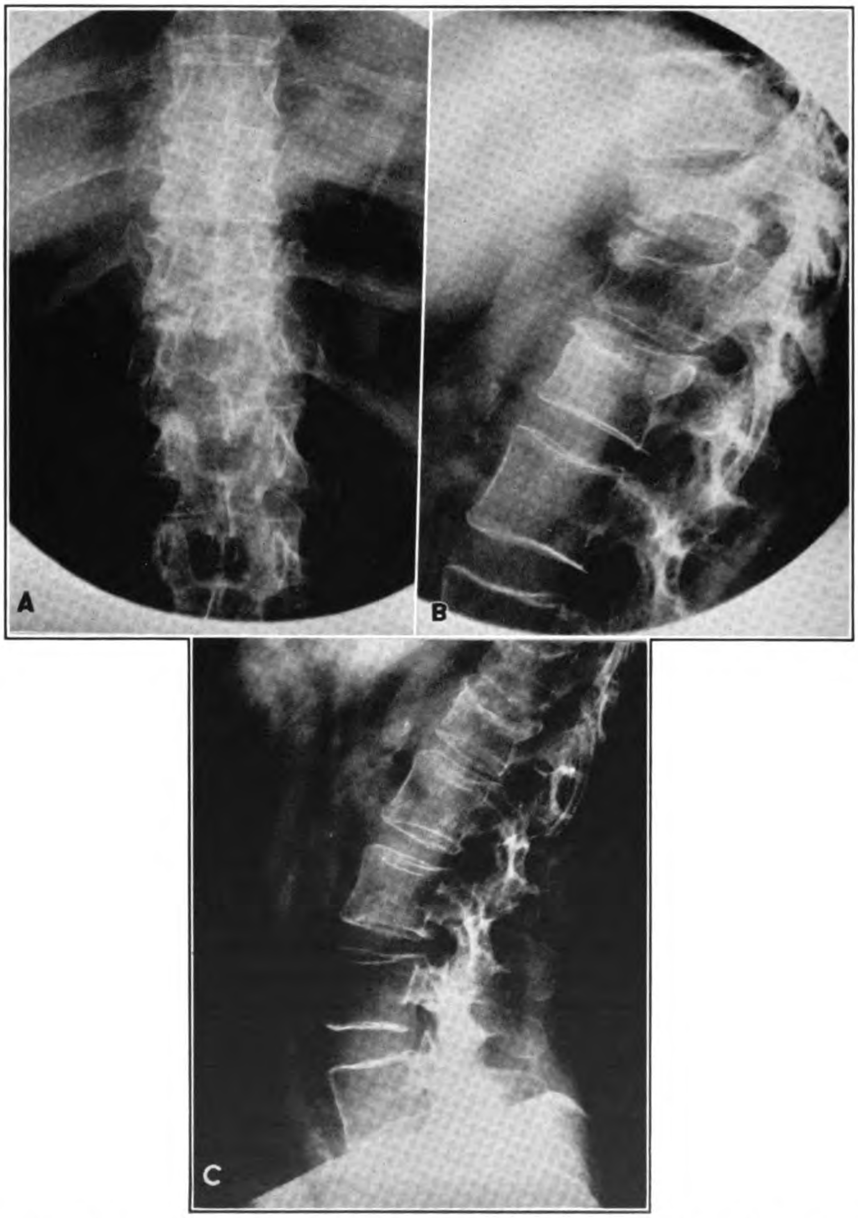

FIG. 114.—Cushing's syndrome in a 37 year old woman. Compression of the twelfth thoracic and first lumbar vertebræ, with slight sclerotic changes, together with marked osteoporosis. The lumbar vertebræ are intact, and the intervertebral discs are somewhat widened but not grossly bulging. Note the sclerotic appearance of the peripheries of the lumbar vertebral bodies.

indicate that "collapse" of the osteoporotic bone structure may take place quietly and continuously, but that this process can sometimes be interrupted by a sudden and more complete compression fracture."

In the first edition of this book it was noted that occasionally osteoporotic vertebræ in patients with Cushing's disease presented compaction of the vertebral surfaces. This was attributed to compression fracture. Incidentally, an illustration of this was published by Sussman and Copleman (1492), but no specific note of its presence was made. In reviewing 12 cases of Cushing's disease from my own experience, this change was found in 3. It appears as a rather broad, band-like zone of increased density which merges rather gradually with the subjacent cancellous bone.

Howland, Pugh and Sprague (1958) suggested that this compaction of the upper, and occasionally of the lower surfaces of the vertebræ may have almost specific importance in the diagnosis of Cushing's syndrome. They found it present in 29 out of 69 cases, occurring only when osteoporosis was more than minimal. As a rule more than one vertebra was affected. The lower thoracic and upper lumbar areas were involved most often, usually together with compression of the vertebræ. In a few cases this change was minimal and its recognition difficult. Posttreatment films after more than six months were available in 19 cases. Eight of these had marginal compressions. This remained unchanged in 2, decreased in 3 and in the remaining 3 had disappeared completely. The condensation was noted to have diminished or disappeared before any change in the osteoporosis was demonstrable radiographically.

Howland, Pugh and Sprague attributed marginal condensation to the effect of the abnormal corticosteroids on the healing of the collapsed portions of bone. Their postulation of a continued strong stimulus for the production of osteoid in Cushing's disease is open to question. In their opinion this strong stimulus for osteoid formation resulted in unusually heavy callus about areas of fracture, and they attributed the heavy callus formation seen about rib and pelvic fractures in Cushing's disease to this mechanism. This does not take into account that osteoid formation and osteoblastic activity is diminished in Cushing's disease. In Sisson's figures 4 and 5 are excellent illustrations of compacted bony trabeculæ in the depressed superior surface of a lumbar vertebra which looks very much like those seen with marginal compression. It is worth recalling his observation about the minimal tissue reaction about fractured trabeculæ, even though in one instance he observed newly formed bone producing thicker structures in the upper surface of a partly collapsed vertebral body (his Figure 18). Insofar as heavy callus about rib and pelvic fractures is concerned, his illustration of a rib with two fractures is illuminating (Figure 19) One of these had neither displacement nor periosteal callus, while the other was surrounded by periosteal callus.

As Howland, Pugh and Sprague point out, marginal condensation, as they term this change, may have specific diagnostic importance in the radiological diagnosis of Cushing's disease. On the basis of present evidence it appears that trauma is a major factor in the production of this interesting phenomenon. However, the mechanism may have some relation to deranged corticosteroid function, and still requires clarification.

With healing of Cushing's syndrome a reversibility of osteoporosis and return of the vertebræ to an approximately normal appearance has been noted in some patients still in the growth period. In others a central zone of osteoporosis persists. As normal bone is laid down at the upper and lower vertebral surfaces the demineralized area becomes sharply delineated. In some the bulging of the intervertebral discs persists, while in others it disappears. Once osteoporosis develops in the postpubertal patient it remains relatively unchanged even when the patient recovers (Iannaccone, Gabrilove, Brahms and Soffer, 1960).

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Acromegaly

This condition is due to eosinophilic adenomas of the pituitary gland. In its fully developed form it is characterized by facial changes, notably a gross coarse thickening of the face and nose, prominence of the jaws, narrowing of the palpebral fissures, and in many cases, prognathism. The concomitant changes in the hands are a marked thickening and enlargement together with a prominent puffing of the ungual tips of the fingers. Less well known are prominent widening and beading of the distal tips of the upper ribs which has been termed "acromegalic rosary."

The accelerated growth is due to an excessive amount of growth hormone. When this occurs before puberty, gigantism results. After the epiphyseal cartilages are united the process of enchondral growth continues at the costochondral junctions, so that the ribs and costal cartilages increase in size and length, contributing to the dorsal kyphosis. Similar changes take place in other areas such as the mandibular condyles. The hormonal effect is not limited to growing bone, but also influences the growth of tendons, fascia, ligaments and joint linings among other tissues.

Changes in the spinal column associated with acromegaly were described by Erdheim in 1931. He pointed out that the vertebral bodies were increased in their anteroposterior dimensions with relatively little increase in their height. This increase in width develops from the periosteum by endochondral ossification of proliferating cartilage. The newly formed bone can be identified roentgenologically by a thin line of demarcation from the vertebral body's original margins. This proliferation of bone is most marked in the anterior aspect of the bone but may occur on the lateral aspects as well. Erdheim pointed out that the new cortex is pathologically wide and that the increased transverse diameter of the vertebral bodies, which is most prominent in the lower thoracic spine may give the mistaken impression of reduced height. The additional intervertebral disc fibrocartilage arises from the perichondrium and is well differentiated into the various normal components including a calcified layer and the hyaline plate. The intervertebral discs retain their normal height.

Acromegalic changes in the spine were reviewed by Chester and Chester (1940) who reported 8 additional cases of their own, 5 of which showed increase in the anteroposterior diameter of the dorsal vertebræ. Waine, Bennett and Bauer (1945) had the opportunity to study at necropsy the spine of an acromegalic patient who died of bacterial endocarditis. They observed that the dorsal vertebral bodies and intervertebral articulations showed massive spur formation with prominent lateral and anterior lipping of the vertebral bodies at the margins of the disc. The histologic picture resembled that of a severe degenerative bone disease. They regarded this as an overgrowth of cartilage and bone secondary to the result of a specific hormonal stimulus.

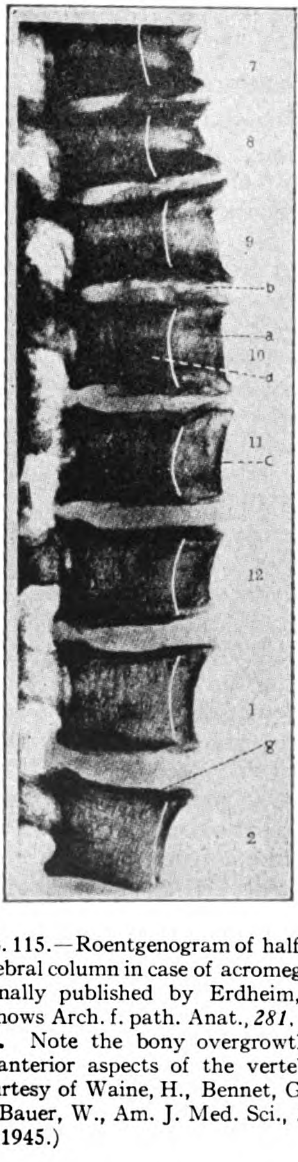

FIG. 115.—Roentgenogram of half the vertebral column in case of acromegaly, originally published by Erdheim, J., Virchows Arch. f. path. Anat., *281*, 197, 1931. Note the bony overgrowth of the anterior aspects of the vertebræ. (Courtesy of Waine, H., Bennet, G. A. and Bauer, W., Am. J. Med. Sci., *209*, 671, 1945.)

Albright and Reifenstein (1948) were of the opinion that the bony changes in acromegaly were chiefly osteoporotic, and might best be ascribed to the hypogonadism which accompanies most of these cases. In 2 cases seen by them in an eighteen-year-old girl and an eighteen-year-old boy the spine showed only osteoporotic changes and scoliosis, with nuclear protrusions into the vertebral bodies. No bony overgrowth was present. They suggested that if this was the correct explanation the condition should respond to estrogen therapy, and reported 2 cases in which this was accomplished. They pointed out that after closure of the epiphyses there was still possibility for restricted endochondral bone formation at certain sites, particularly the cartilaginous end plates of the vertebræ. These may again start producing endochondral bone, which together with increased periosteal bone formation accounts for the increased width of the vertebræ in the anteroposterior diameter.

The question arises as to the relative incidence of changes in the vertebral column associated with acromegaly. We reviewed the available films of the last of 23 cases of acromegaly seen by us without finding any radiologic evidence of gross vertebral changes in any of them. This indicates that the change is not one which commonly accompanies acromegaly, but in no way negates the positive evidence adduced by other authors. The reader is directed to the illustrations published by Waine, Bennett and Bauer (1945), Gilmore and Mahan (1947), and Steinbach, Feldman and Goldman (1959) as excellent pictures of this condition.

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HYPOTHYROIDISM

This condition is incident to a diminution in the secretion of thyroid hormone. It may be partial, or if the gland is completely absent either as a result of destruction or surgical removal, a state of athyrosis may exist. Hypothyroidism may be encountered in any age group from infancy to adult life. Hypothyroidism originating in infancy or during fetal life is termed cretinism, and may be recognized by thickening of the subcutaneous tissues and the characteristic facies with a dull skin, apathetic expression, enlarged and often protruding tongue, thick lips and a wide flat nose. These children also may develop a characteristic protuberant abdomen with short arms and legs. Mental deficiency is common and varies from an almost complete imbecile state to the level of a moron. Growth in these children is retarded and the osseous development may be considerably delayed.

From the radiologic viewpoint these children present evidence of dysgenesis of the epiphyses of the wrists, elbows and knees (Wilkins, 1941). Development of dentition is frequently retarded with delayed eruption of the teeth. Bony changes observed *in utero* were reported by Dorff (1934).

The radiologic changes in the spine reflect the delayed development of the skeletal structures. The gibbus deformity described below apparently may recede under thyroid treatment (Evans, 1952). The neurocentral synchondroses may have delayed union. The vertebral bodies often show some degree of osteoporosis. Engeset, Imerslund and Blystad (1951) described fragments of irregularly ossified centers within the vertebral bodies as well as evidence of arrested longitudinal growth. In one of their cases they also reported a kyphotic angle at the first and second lumbar vertebræ similar to the findings noted in Hurler's disease. They suggested that this change in the appearance of the anterior aspect of the upper lumbar segments might perhaps be found more often if searched for. Swoboda (1950) reported 7 cases of infantile myxedema, all of which had incomplete wedging of the second lumbar vertebra producing an angular kyphosis. This was variable in degree, and in most of his cases produced a clinical gibbus. The involved vertebral bodies had a step-like wedging anteriorly and posteriorly. Caffey (1956) mentioned a case of hypothyroidism in a boy three years of age in whom the maturation of the vertebræ was retarded. The first lumbar body was hypoplastic, producing a kyphotic deformity which persisted despite otherwise effective thyroid treatment. Nine years later the patient still had a marked kyphosis with spondylolisthesis and vertebral deformity. Another case of his in an untreated hypothyroid girl eight years of age revealed opened neurocentral synchondroses together with an oval anterior notching in the vertebral bodies characteristic of the first year of life. Evans (1952), examined 13 cases of cretinism, and found minor abnormalities in 5. The other 8 all had deformities of the bodies of the twelfth dorsal or first and second lumbar vertebræ with kyphotic changes. Similarity to the changes present in achondroplasia and gargoylism was pointed out.

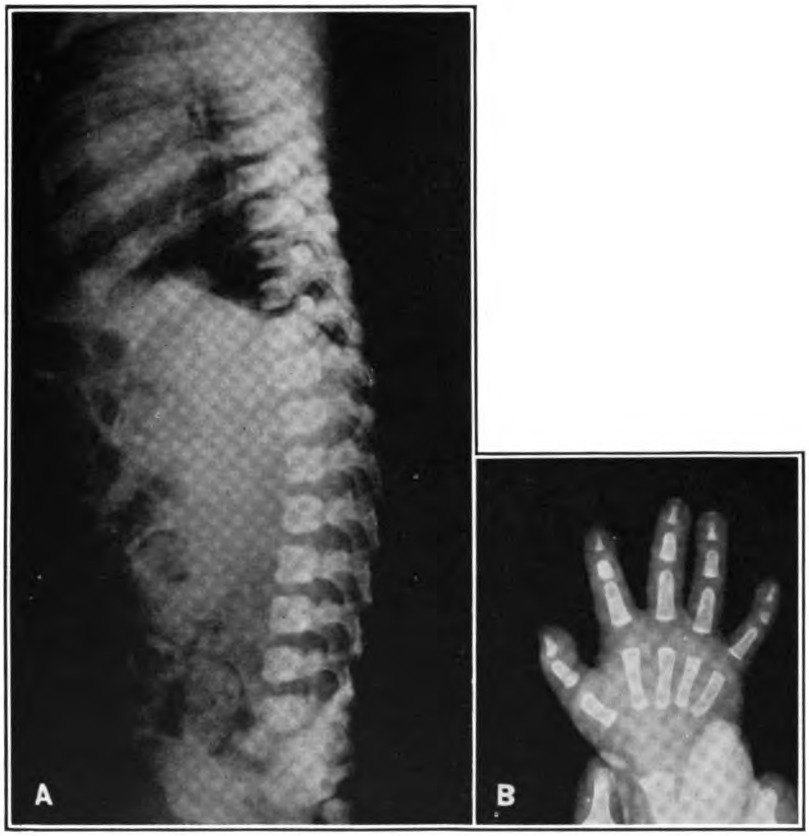

FIG. 116.—*A*, Hypothyroidism in a nineteen-month-old child. Note the relatively increased densities of the upper and lower vertebral plates and the central area of diminished density. *B*, Same case. Retarded epiphyseal development of the hand and wrist.

In our own cases of cretinism retardation of skeletal maturation was evident, as reported by many others. Studies of the vertebral column in these patients disclosed a slight degree of osteoporosis of the vertebral bodies. This was not considered a distinguishing feature of the disease. Neither was there any definite change in the configuration of the vertebral bodies as described by other authors, which merely indicates that we still have to look for these changes.

In 1 case of hypothyrosis under our observation an atypical configuration of vertebral bodies was noted. There was a definitely increased density of the superior and inferior aspects of the various vertebræ, particularly in the dorsal region between which a rarefied zone could be observed. This condition had been well described by Engeset and his co-workers (1951) who described a persistent center of osteoporosis persisting after actual bone growth has become normal as indicative of the fact that the old bone remains abnormal, while new bone is laid down with normal density. They commented on these changes as being somewhat similar to those seen with scurvy.

Another change associated with hypothyroidism in children is a dysgenesis in ossification centers such as the femoral head, the distal epiphyses of the femurs and the wrists, producing a stippled configuration sometimes associated with a flattening of the affected epiphyses. Under adequate thyroid treatment these changes disappear and growth proceeds normally in successful cases. This type of stippling has rarely been described in the spinal column. With healing of the hypothyroid state the signs of epiphyseal dysgenesis in the thoracolumbar spine tend to diminish (Middlemass, 1959).

The condition known as juvenile myxedema refers to hypothyroidism occurring in a previously normal child before the onset of puberty. In these cases some degree of delayed ossification occurs, together with diminution in growth.

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TUBEROUS SCLEROSIS

This condition is considered a congenital hamartial ectodermoses, one of a group which includes von Recklinghausen's neurofibromatosis, Sturge-Weber syndrome and Hippel-Lindau's disease. The underlying tumor is a potato-like growth which varies in size from 0.5 to 3.0 cm. in diameter. These occur most frequently in the cerebral hemispheres, although other parts of the central nervous system may be involved. The masses may project into the ventricles from immediately beneath the ependyma, and calcification may occur within these tumor masses. Microscopically the pyramidal cells and glia are most affected. The condition is associated clinically with epilepsy, mental deficiency, adenoma sebaceum and congenital tumors of the eye and many visceral organs. In some cases the endocranial lesions may dominate the picture, while in others the visceral tumors are responsible for the patient's complaints. Involvement of the skeleton is relatively infrequent.

Radiologic changes, aside from intracranial calcifications, include diffuse and widely varying skeletal manifestations of cystic, sclerotic or combined such lesions. These occur as periosteal thickening, fragmentation of cortical layers of bone, mottling of the skull of a sclerotic nature, rarefied changes in the skull, and cyst formations particularly in the terminal phalanges.

The vertebræ may contain multiple sclerotic foci which are scattered through the whole of the spine and pelvis, including the neural arches. These can be confused with osteoplastic metastatic lesions. Occasionally a cystic rarefaction appears in a vertebral body.

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PROGERIA

This is a condition of premature old age and represents a rare form of senilism which has a rapid onset, with retention of normal intelligence and without a perceptible cause. The condition is marked by a loss of hair, shrivelling of the nails, congenital cataracts and emaciation, together with a loss of subcutaneous fat and precocious arteriosclerosis (Moehleg, 1946). Roentgenologic examinations usually reveal a diffuse osteoporosis with swelling of the epiphyses and no change in the centers of ossification. Scleroderma together with hypoplasia of the terminal phalanges of the hands and feet may occur. The skull may be rather large with slightly separated suture lines. Very little is mentioned about the spine in these rare cases. Talbot, Butler, Pratt, MacLachlan and Tannheimer (1945) reported a case in which they had carried out metabolic studies during the year preceding death from coronary thrombosis at seven-and-one-half years in a case of progeria. Roentgenograms of the spine in this patient showed an unusual degree of anterior notching in the lateral views. At necropsy no cause for the condition could be ascertained.

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HYPOPHOSPHATASIA

This is a chronic, familial condition characterized mainly by rachitic-like deformities in children, associated with low serum and tissue alkaline phosphatase activity and defective bone formation. The disease may begin *in utero*, and manifests itself in neonatal life or later. In the newborn severe deformities of the extremities are present, together with soft skeletal structures and a globular skull which has markedly deficient ossification. The calcification of the skeleton likewise is scanty, and there is severe deficiency in the articular structures, the ends of the bones showing widely separated areas corresponding to the bony deficiencies. Multiple fractures occur. Infants so afflicted rarely survive.

When the disease appears after an initial period of apparent normalcy, symptoms of anorexia, vomiting, hypotonia and failure to thrive develop, together with cyanosis and convulsions. In these patients the joint structures are prominent, the skull enlarged and rachitic changes are present in the epiphyses and costochondral junctions. If the patient survives the skeletal changes interfere with walking and standing, and craniostenosis appears. In children in whom the onset of hypophosphatasia takes place after about 18 months of age similar bony changes are present, and the development of the child is retarded. Craniostenosis is less frequent as a later manifestation. Dentition is deficient in both groups. The pathologic changes have been described as resembling osteogenesis imperfecta in the newborn, while in older patients there is resemblance to rickets. The cause of death is usually renal failure. In those who survive residual and sometimes prominent skeletal deformities persist.

The vertebral column of newborns with hypophosphatasia partakes in the marked failure of ossification. In those who survive deficient calcification is manifested by delayed and irregular bone formation, while still later relatively minor vertebral malformations with intact bodies and occasionally dorsolumbar angulations are present.

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METABOLIC DISORDERS

Osteoporosis

In osteoporosis the decrease in bone tissue is ascribed to a disorder of tissue metabolism characterized by the failure of the osteoblasts to lay down bone matrix. Inasmuch as this is a disease of tissue metabolism, and not of calcium metabolism, it is associated with relatively normal serum calcium and phosphorus levels. The most common cause for osteoporosis in the adult is the postmenopausal state caused by a deficiency in estrogen and consequent failure of adequate osteoblastic activity (Albright, Smith and Richardson, 1941). Probably next frequent is the so-called senile atrophy in which the bony structures undergo the same type of atrophy as all of the other components of the body. The atrophy of disuse with diminished osteoblastic activity, the so-called idiopathic osteoporosis of unknown etiology and the osteoporosis of malnutrition with depletion of bone matrix because of insufficient protein intake are among the other causes which have to be kept in mind. Among the glandular causes, some of which already have been discussed, are Cushing's syndrome, the adaptation syndrome of Selye, acromegaly, primary ovarian insufficiency (Albright, Smith and Fraser, 1942) and congenital osteoblastic defect as exemplified by osteogenesis imperfecta. A second major group included defects in bony matrix due to loss of androgen such as is seen with eunuchoidism and possibly senile osteoporosis (Albright and Reifenstein, 1948). An interesting case of primary ovarian agenesis in a seventeen-year-old girl was reported by Radwin, Michelson and Shore (1949). I have observed a girl of thirteen years presumed to have a pituitary adenoma with spinal osteoporosis. Other conditions associated with osteoporosis in relatively rare instances include polycythemia vera, myesthenia gravis, Gaucher's disease and Waldenstrom's macroglobulinemia. Diffuse, and often extensive demineralization of bone is noted frequently in children with leukemia, either along or together with the effects of steroids. The more advanced changes usually follow both the concomitant course of the disease and treatment. The demineralization of bone associated with metastatic malignant disease is well known. Its origin also includes factors such as disuse, postmenopausal hormonal alterations and probably other still unknown factors. Ankylosing spondylitis also is frequently accompanied by pronounced osteoporosis which may be heightened by steroids and disuse.

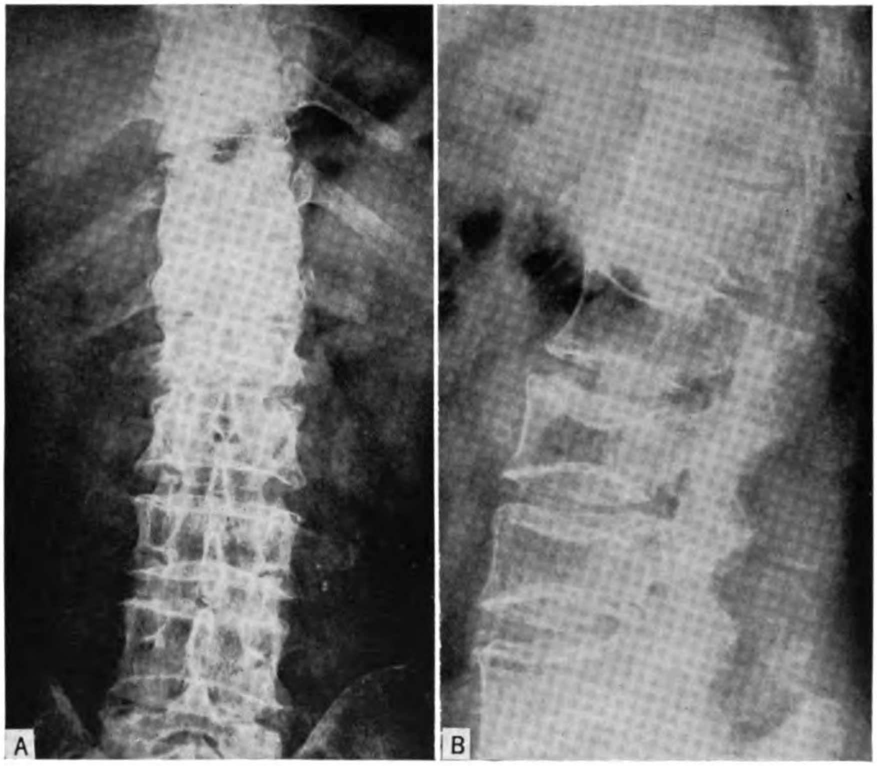

FIG. 117.—Osteoporosis, postmenopausal, in a 58-year-old woman with intermittent midback pain. The twelfth thoracic and second lumbar vertebræ are compressed. The intervertebral discs are biconvex and are quite prominent, indenting into the articular surfaces of the respective vertebral bodies. The cancellous bone is diminished, the cortical bone is thinned but relatively dense.

Three factors enter into the decreased activity of the osteoblast (Albright, 1947). These are steroid hormones, mechanical stresses and strains, and nitrogen building blocks. With respect to the bone matrix, steroids are divided into anabolic or "N" or "nitrogen" hormones, (estrogens and androgens), anti-anabolic (adrenocortical "sugar" or "S" hormone and anabolically inert steroids (progesterone). The "N" hormone is opposite in action to the "S" hormone. The osteoporosis of old age is attributed in part to the loss of gonadal hormone and of adrenocortical "N" hormone, and responds to estrogen and testosterone treatment. Cushing's syndrome and Selye's adaptation syndrome are considered to be due to an excess of anti-anabolic adrenocortical "S" hormone and this, too, responds to testosterone treatment. The adreno-corticotropic hormone of the pituitary by releasing the "S" hormone from the adrenal cortex likewise produces osteoporosis (Albright, 1947).

Cocchi (1951) described a form of hepatogenic osteoporosis which occurs when there is extensive destruction of liver parenchyma, such as may result from toxic influences, biliary occlusion or external bile fistulas. Similar changes may be encountered in cirrhosis or in extensive liver metastases.

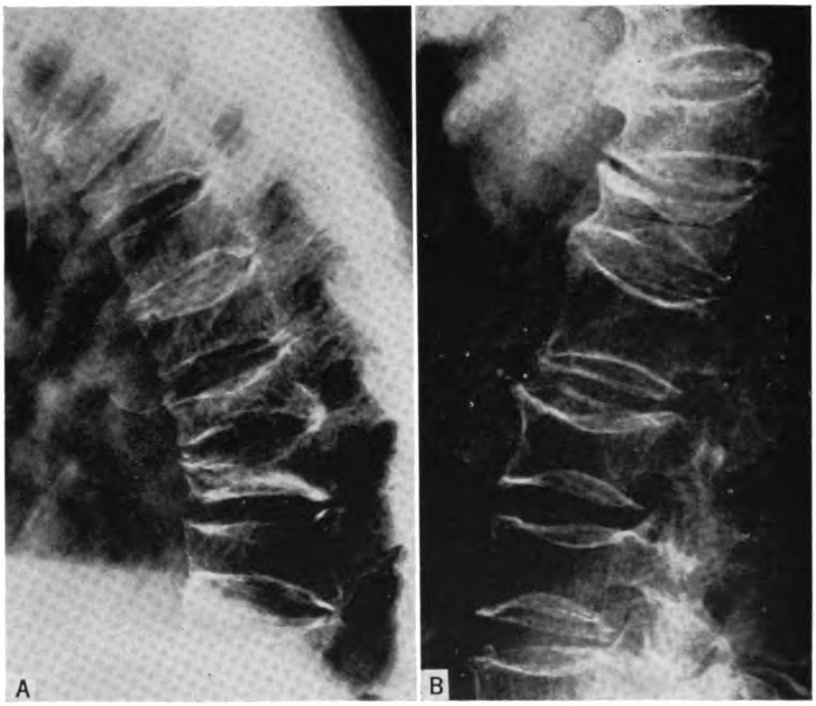

FIG. 118.—Thoracic and lumbar spine, lateral projection, in a 65 year old man with senile osteoporosis. There are multiple collapsed vertebral bodies. The intervertebral discs are variably widened. Some condensation of bone is noted along the inferior aspect of the collapsed ninth thoracic vertebra (*A*). Note the relatively increased density of the vertebral peripheries (*B*). The vertical trabeculæ are better seen in the lower lumbar area (*B*).

The mechanism for this change is complex, including the disturbance of calcium and fat resorption from the intestine and faulty absorption and deposition of vitamin D in the liver. Among the other findings were secondary hyperparathyroidism due to lack of calcium, atrophy of the testes due to failure of the liver's inactivating functions on estrogens, and hyperplasia of the adrenal cortex due to atrophy of the testes resulting in overproduction of the adrenocortical "S" hormone.

The association of hyperthyroidism with osteoporosis was exemplified in a case reported by Flink (1948) in a sixty-four-year-old woman with marked osteoporosis, particularly of the spinal column. This was ascribed to increased excretion of calcium and phosphorus in the urine and feces, the continuous negative calcium balance manifesting itself eventually as osteoporosis. Instances of juvenile hyperthyroidism with osteoporosis were described by Jacobs (1943).

Osteoporosis with ulcerative colitis was described by Ricketts, Benditt and Palmer (1945). Miyakawa and Stearns (1942) reported a case of severe osteoporosis or osteomalacia in a thirty-four-year-old woman who had had steatorrhea for more than ten years.

The bony changes associated with osteoporosis of whatever cause are alike. Microscopic examination of the bone shows the spongiosa and cortex to be thin, but to consist of normal lime-containing osseous tissue. The trabeculæ of cancellous bone are widely separated and red bone marrow may be considerably diminished with replacement of a greater part of the bone marrow by fatty material.

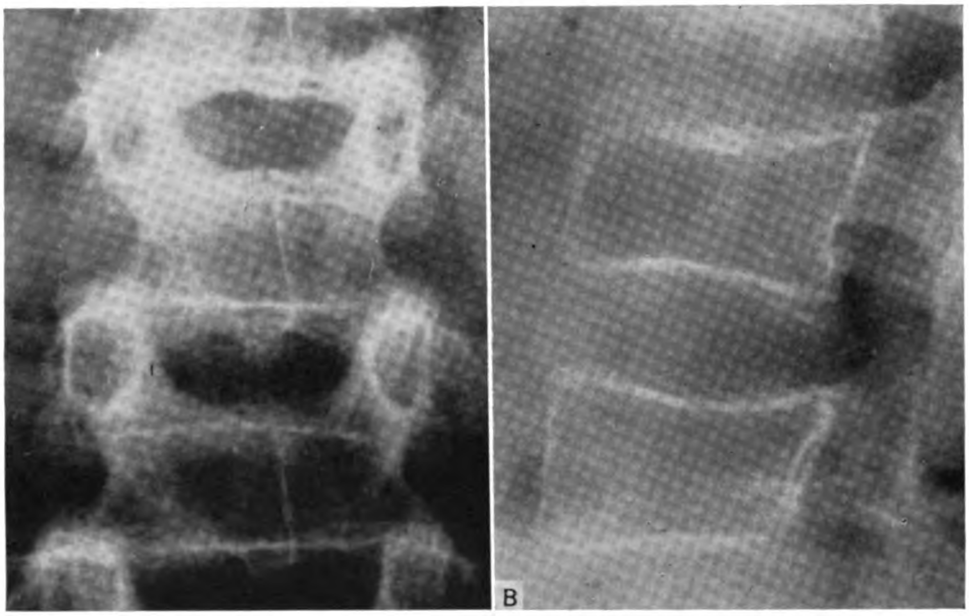

FIG. 119.—Anteroposterior and lateral roentgenograms of upper two lumbar vertebræ of a 16-year-old boy with iatrogenic Cushing's syndrome due to prolonged intake of cortisone for chronic skin malady. The intervertebral discs are ballooned. The vertebral bodies are biconvex, with relatively heavy but thinned cortical borders. The trabeculations are reduced, the vertical components are being somewhat better maintained.

Cocchi (1951) reported that in his case histologic examination of the bone showed simple osteoblastic or osteoclastic osteoporosis or bone absorption, with and without osteoid bands. Snapper (1952) described bone changes associated with parathyroid hyperplasia in experimental animals with a biliary fistula or chronic jaundice. Bone changes in humans with such fistulæ are rare. Snapper mentioned that his patients who developed bone lesions of clinical importance under the influence of a biliary fistula or chronic jaundice were old women, in whom the possibility of senile osteoporosis or postmenopaural osteoporosis could not be excluded. He included the complete report of a case of Hitzig in whom chronic obstructive jaundice was accompanied by a severe form of osteoporosis with vertebral compression in a forty-eight-year-old woman.

Among their cases of so-called idiopathic osteoporosis Albright and Reifenstein included two young women in both of whom osteoporosis was made worse by pregnancy.

The production of osteoporosis by the prolonged use of cortisone, hydrocortisone and corticotropin was reported by Demartini, Grokoest and Ragan (1952) and by Boland (1952). Such treatment resulted in clinical features simulating Cushing's syndrome if enough material is used to produce hyperadrenalism. In the group reported by Demartini, Grokoest and Ragan 5 patients had been treated from five to twenty months. All were women, and 4 were in the postmenopausal age group. A predisposing factor such as prolonged bed rest, prolonged activity of the disease under treatment or postmenopausal states were present in each patient. Boland stated that 7 of his patients had spontaneous fractures. Spinal compression fractures appeared in 4, fracture of the femoral neck appeared in 2 and a pubic ramus fracture occurred in 1. Each of his patients had had long-standing rheumatoid arthritis. Osteoporosis and pathologic fractures in patients treated with corticosteroid therapy over a prolonged period of two and a half years for pemphigus was reported by Teicher and Nelson (1952).

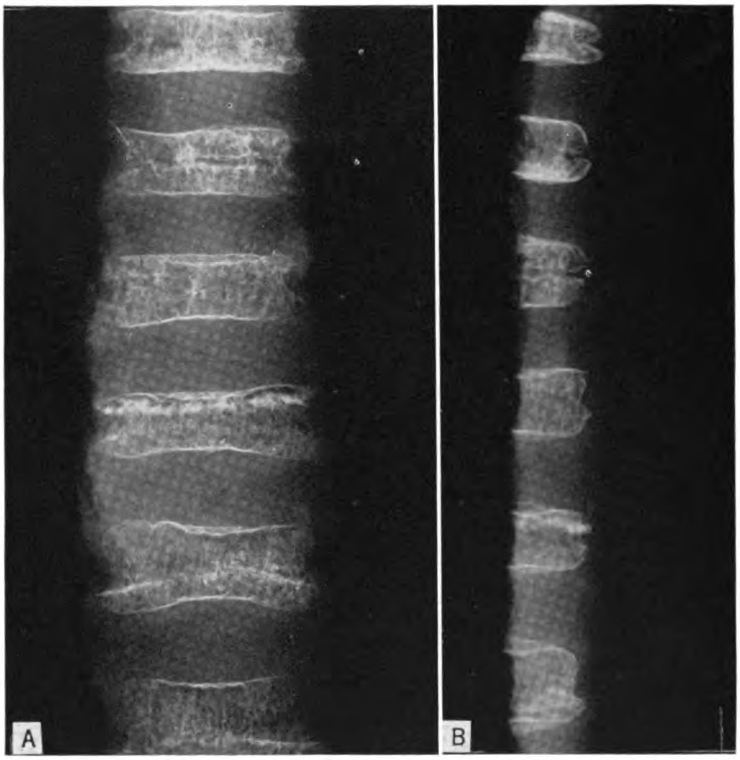

Fig. 120.—Anteroposterior and lateral roentgenograms of slice of thoracolumbar spine of infant with severe osteoporosis associated with congenital biliary atresia. The vertical trabeculæ are better maintained. Note the compaction of the trabeculæ in the superior aspect of the vertebra fourth from the top of this illustration.

When osteoporosis has advanced to a certain degree the trabeculæ are no longer able to resist the weight of the body and the internal pressure of the intervertebral discs. This results in collapse of the central portion of the vertebral body producing a biconcave vertebra and expansion of the nucleus pulposus and disc in a biconvex fashion follows.

Radiologic examination of the spine in early cases of osteoporosis first reveals a diffuse demineralization of the vertebral bodies. At this stage the vertebral bodies retain their normal contours, but present a glazed appearance with rather sharply marked borders of increased density at the periphery of the respective vertebræ. The intervertebral spaces as a rule are not disturbed. At this stage it is not uncommon for impaction fractures to occur in the anterosuperior aspect of the vertebral bodies, especially in the mid-thoracic and the thoracolumbar regions. The presence of Schmorl's nodes may also be observed, but of interest is the fact that sclerosis of the bone surrounding the herniated nodules of fibrocartilage is conspicuous by its absence. With further progression of the osteoporotic process the demineralization increases and the tensile strength of the vertebræ becomes lessened. This may be manifested by a diffuse collapse of one or several vertebral bodies, particularly in the mid-thoracic and upper lumbar regions, together with associated widening of the intervertebral spaces. Anterior wedging of the vertebral bodies with concomitant dorsum rotundum may become prominent. In the advanced stages of osteoporosis the vertebral bodies may assume a biconcave configuration of the intervening intervertebral cartilages. Combinations of diffuse flattening of the vertebral bodies together with biconvex configurations may be observed, with flattened vertical bodies more prominent in the thoracic region.

Involvement of the cervical spine by osteoporosis may result in demineralization of the vertebral bodies. Osteoporotic collapse of the cervical segments is uncommon. The cervical intervertebral discs likewise usually maintain normal contours unless altered by discogenic disease, which is frequent in the age group in which osteoporosis is most common.

The remainder of the skeleton usually shows porotic changes of lesser degree, but in advanced cases thinning of the cortices of the long bones and widening of their medullary canals may be observed fairly often. A change which has recently been associated with osteoporosis particularly of the postmenopausal, senile or idiopathic type, is parietal thinness of the skull bones (Epstein, 1952).

The osteoporosis of the vertebral column from the causes noted above may bear a marked similarity to that associated with other diseases, particularly osteomalacia, myeloma, certain forms of hyperparathyroidism and the equally interesting but less common conditions mentioned before. These may be distingusihed by considering the clinical pictures associated with them in differential diagnosis.

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Osteomalacia

This condition refers to disorders which result in the presence of abnormal amounts of osteoid tissue in the bones because of insufficient calcification. Any condition which depletes the body's calcium stores sufficiently can cause osteomalacia. Such depletion may take place through insufficient absorption of calcium or through excessive loss. Insufficient quantities of inorganic phosphorus in the blood is another factor which militates against normal calcification of bony matrix by whatever calcium is present in the serum. Blood chemistry reveals a serum calcium value which is within normal limits or somewhat diminished. The blood serum phosphorous level likewise may be normal or slightly lower than usual. The failure of bone to calcify properly leads to increased osteoblastic activity with a consequent elevation in the serum alkaline phosphatase content. Albright and his co-workers indicated that rickets presented all the characteristics of osteomalacia together with additional changes at the epiphyseal line, principally faulty calcification of the zone of provisional calcification (failure of osteoid tissue to calcify normally).

Albright and Reifenstein separated osteomalacia into four categories depending upon the degree of severity. The first group included chemical osteomalacia with normal phosphatase, the second chemical osteomalacia with high phosphatase, the third Milkman's syndrome and the fourth advanced osteomalacia. The degree of severity is dependent upon the relationship between serum calcium and serum phosphorus content. When the change is minimal the disordered deposit of calcium and osteoid tissue is not sufficient to give rise to sufficient skeletal weakness to produce a high phosphatase content because of increased osteoblastic activity. When the osteoblastic activity is stimulated, and where radiologic and clinical indications of osteomalacia are lacking but still phosphatase content is elevated, there exists a group of cases in which neither pseudo-fractures nor demineralization occur. Cases with chemical osteomalacia and pseudo-fractures without obvious generalized demineralization constitute the third group, of which Milkman's syndrome is probably the best example. The last classification refers to those people with an advanced softening of bones characteristic of osteomalacia.

From an etiologic viewpoint Albright and his co-workers pointed out that osteomalacia due to simple vitamin D deficiency has not been observed in this country, but has been reported by Maxwell (1935) as occurring frequently in Northern China. Recently Winston and Pendergrass (1954) reported a case of Milkman's disease which they attributed to simple vitamin D deficiency in an adult. Vitamin D resistant rickets as seen in clinical practice in this country as well as elsewhere, and steatorrhea, are other causes. The next group described by these authors is due to renal acidosis as caused by the Fanconi syndrome and the renal disease of tubular insufficiency without glomerular insufficiency. A third group is idiopathic hypercalciuria and a fourth is hyperparathyroidism with osteitis fibrosa generalisata during the transitional stage following the removal of a parathyroid tumor.

There are also bizarre metabolic diseases in which osteomalacia plays a significant role. For example, Lowe (1952) mentioned marked bone loss and suggestive rachitic changes as occurring in an unusual syndrome consisting of organic acidemia, diminished renal ammonium production, hydrophthalmos and mental retardation. In one such case seen by us recently the rachitic-like disturbances responded to vigorous vitamin D therapy, but the osteomalacia, particularly of the spine, did not recede visibly. The syndrome of idiopathic hypercalcuria, caused by impaired tubular reabsorption of calcium, results in increased calcium excretion and nephrolithiasis, also includes a degree of osteomalacia among its manifestations. Evidence also has been presented to indicate that osteoporosis occurring with prolonged corticosteroid treatment may in later stages be accompanied by osteomalacia.

The radiologic features of Milkman's syndrome have received considerable attention. At first (Milkman, 1934) this was considered to be a specific disease of unknown etiology, but the work of Albright and his colleagues (1946) among others has resulted in the inclusion of this syndrome in the group of osteomalacic diseases. The condition is characterized by multiple spontaneous idiopathic symmetrical pseudo-fractures. These zones of pseudo-fracture are actually united, the symmetrical focal bands of translucency being due to zones of osteoid tissue. The outstanding radiologic feature consists of these translucent band-like areas. At first no deformity is present but later separation, displacement and overriding of fragments may occur. Settling of the vertebral column, formation of a heart-shaped pelvis, coxa vara, rib, ulnar and clavicular angulations are part of the picture of generalized osteomalacia. Usually no callous formation is observed in these individuals. Albright and his colleagues mentioned that bone tissue taken from a rib and vertebra of 2 patients with Milkman's disease showed characteristic changes of osteomalacia. They favored the retention of Milkman's syndrome as an entity to emphasize the fact that osteomalacia can be present without generalized decalcification, with only these united uncalcified zones. In their discussion they presented 2 cases of Milkman's syndrome associated with steatorrhea.

In a report of a thirty-seven-year-old male patient with Milkman's syndrome, Brick and Bunch (1947) reported that roentgenograms of the spine showed narrowing of the fifth and sixth dorsal vertebræ, with pseudo-fractures of the spinous processes of the fifth, sixth and seventh cervical vertebræ. There was generalized osteoporosis with pseudo-fractures of the interarticular isthmus of the third, fourth and fifth lumbar segments, of the second to fourth, the sixth, seventh, ninth, tenth and eleventh ribs, the manubrium and other bones. All in all this patient had 39 fractures. Biopsy of the tibia showed fibrosis and thinning of the cortex with unusually large Haversian canals and rarefaction of the bone.

Pseudo-fractures may occur with many diseases including osteomalacia, richets and latent rickets, renal rickets, celiac disease, chronic idiopathic steatorrhea non-tropical sprue, early and late osteogenesis imperfecta, fragilitas osseum, hyperparathyroidism, hyperthyroidism, Paget's disease, adrenal pituitary bone dystrophy, severe chronic acidosis, hyperglycemia, congenital syphilis, osteomyelitis, osteopetrosis, march fractures and bone dysplasias. The common feature to all of these is weakening of the bony framework or excessive load on the bone. Such lesions are found most often in the lower extremities, the pelvis, ribs, forearms, and wrists. In the vertebræ they may be difficult to distinguish from compression fractures (Camp and McCullough, 1941).

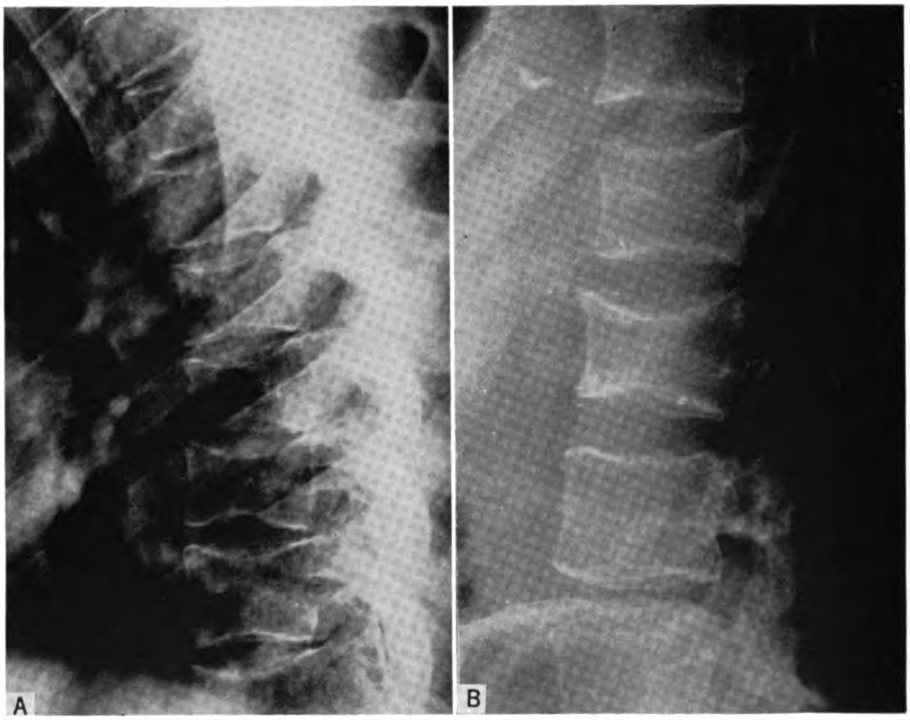

Fig. 121.—Osteoporosis and osteomalacia in a 27-year-old woman complaining of persistent back pain for a month after delivery of a normal infant. The intervertebral discs are bulging in an irregular pattern. Some of the vertebræ are less dense than others, and varying compressive changes are present.

Albright, Burnett, Parson, Reifenstein and Roos (1946) did not entirely agree with the conclusions expressed in the preceding paragraph. They observed that the conditions listed were either rickets or osteomalacia, depending on the age of the patient, and in the others the resemblance was purely superficial. They pointed out that in Paget's disease, polyostotic fibrous dysplasia and osteogenesis imperfecta these fractures occurred through areas of bony pathology, while in osteomalacia they occurred in bones which appear otherwise perfectly normal radiologically.

In our own series we have had several cases of osteomalacic diseases, one of which is reproduced here (Fig. 122). The spine roentgenograms revealed a diffuse osteoporosis interspersed in which were numerous web-like lines

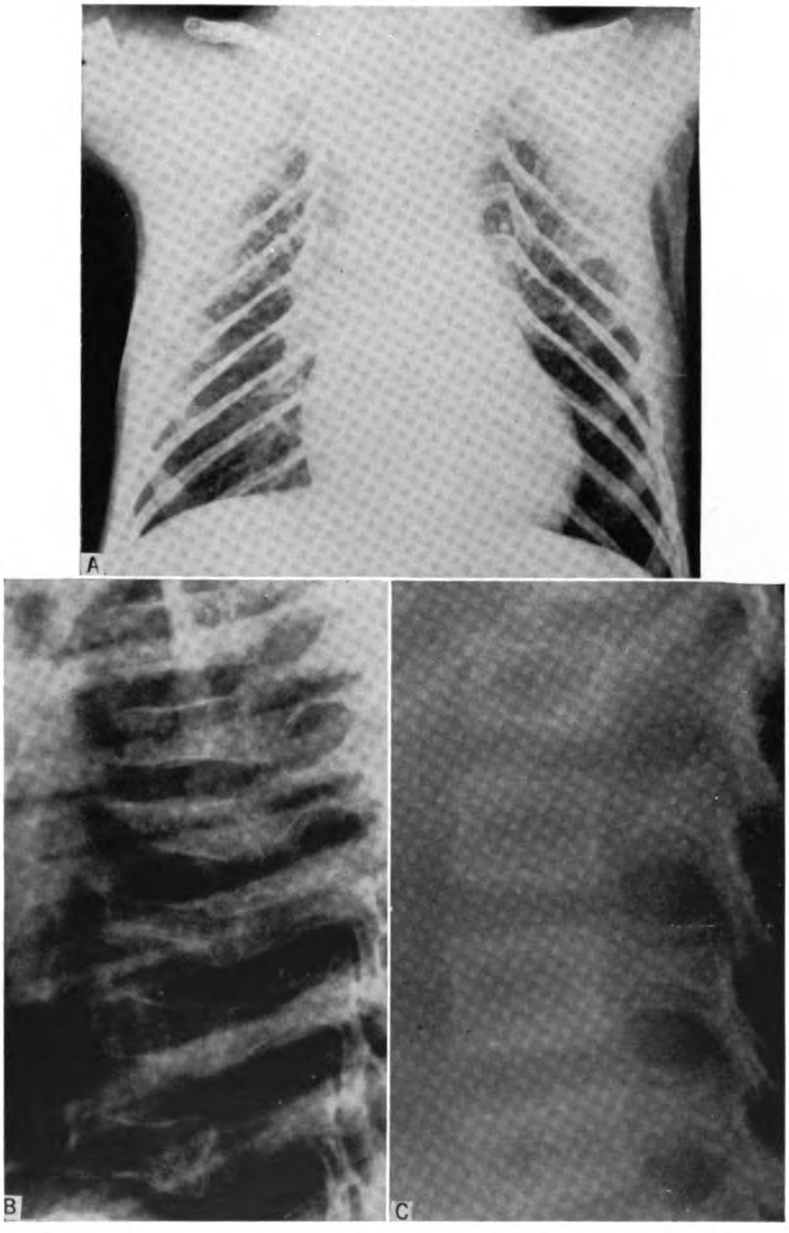

FIG. 122.—Osteomalacia in an eight-year-old boy who had undergone famine about four years preceding this examination. The ribs are slender, and are angulated downwards beginning near the costovertebral articulations. The thoracic vertebræ are diffusely flattened, and the intervertebral discs are slightly biconvex. The lumbar vertebræ are demineralized still, but their contours are well maintained. The lumbar discs are moderately bulging in appearance. The vertebral peripheries are slightly denser than their central portions.

of increased density. Several of the vertebræ in the lower and mid-dorsal spine were reduced in height. Pseudo-fractures could be demonstrated in the inferior aspect of the body of the fourth lumbar vertebra. There were no symptoms referable to the vertebral column changes. The underlying cause was believed nutritional, the child having been exposed to famine.

A case observed by us was considered as having both osteoporosis and osteomalacia on a nutritional and biliary basis. This was a sixty-seven-year-old man who following a subtotal gastric resection for a duodenal ulcer was unable to accept food for a long time and developed a severe protein deficiency. A long-draining biliary fistula formed. Radiologically this was accompanied by pronounced osteoporosis resulting in practically complete demineralization of his spine, with biconcave vertebræ and markedly enlarged and biconvex intervertebral discs. This patient also developed a marked degree of decalcification of the remainder of his skeleton. The pelvis was of interest in that the femoral heads and acetabulæ protruded deeply into the true pelvis. His skull presented bilateral parietal thinness. This patient improved slowly but steadily on hormonal therapy together with an enriched diet. During this long period of recovery he fell and fractured his right hip, which was subsequently pinned with a good clinical result. Not noted at the time of the original film was the presence of a definite osteoid zone in the opposite left femoral neck. The patient complained of mild pain in the left hip, but a subsequent radiologic examination failed to disclose any evidence of fracture. About seven months later this zone was recognized in retrospect when a current roentgenogram revealed that the left femoral neck had been fractured without known trauma, the

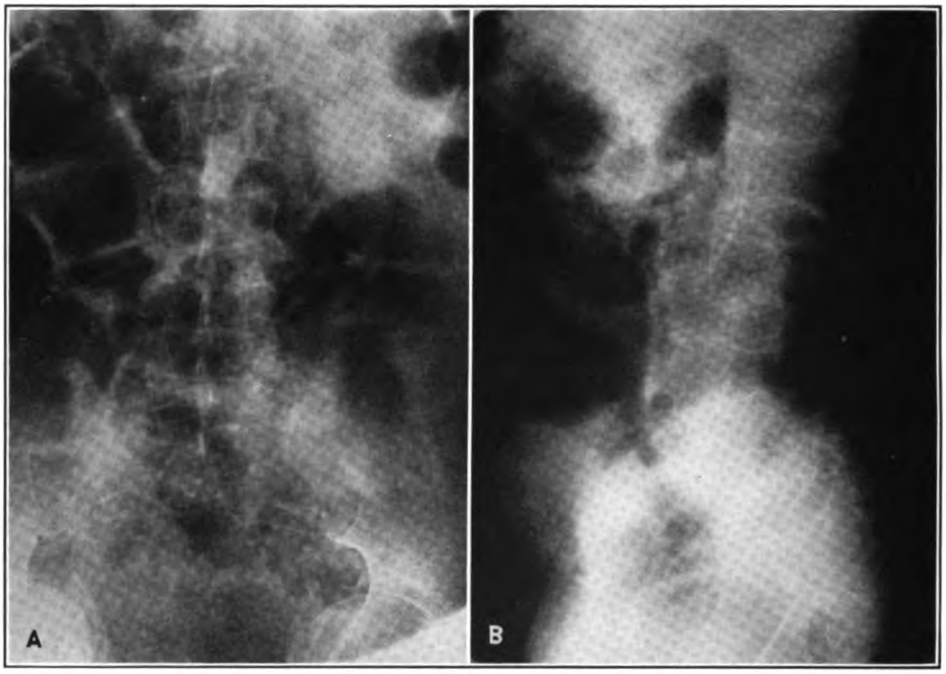

FIG. 123.—Osteomalacia following subtotal gastrectomy for ulcer complicated by draining biliary fistula. Note the advanced demineralization of the vertebræ, the bulging intervertebral discs, and the inward protrusion of the femoral heads, particularly the left one.

fracture line apparently occurring through the osteoid zone. Repeated blood chemistry studies in this patient showed the serum calcium to be between 9 and 10 milligrams per cent, the serum phosphatase to be between 3 and 4 milligrams per cent, and the serum alkaline phosphatase value to be between 10 and 15 milligrams per cent (Fig. 123).

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RICKETS

Infantile rickets associated with vitamin D deficiency is associated with failure of calcification of osteoid tissue and bone development due to an insufficient utilization of calcium. A reduced serum phosphorus content is present, which is caused by failure of absorption of phosphorous from the intestinal tract, and increased excretion of phosphorus from the intestinal and urinary tracts. The failure of osteoid calcification has been mentioned in the section under osteomalacia. The characteristic radiologic alterations in the growing bones include widened epiphyseal zones, with flaring and tuft-like configuration of the metaphyseal ends of the growing bone. The diffuse osteomalacia sometimes associated with rickets may manifest itself in the spine by a similar process. The superior and inferior surfaces of the respective vertebral bodies may present a convex appearance, with the height of the vertebral bodies somewhat reduced and the intervertebral spaces proportionately widened. A mild dorsolumbar kyphosis has been observed in several cases of severe infantile rickets, an example of which is included here. With proper therapy the vertebral bodies regain their height and normal osseous density returns.

Oppenheimer (1939) described rarefaction of the vertebræ in early stages of rickets, with rapid progression. In less acute phases he identified globular rarefied areas arranged like clover leaves which later became surrounded by calcium as the rickets healed. Broadening of the epiphyseal cartilages was not found, but compression of the vertebral bodies and expansion of the discs occurred if rarefaction was sufficiently marked.

Albright, Butler and Bloomberg (1937) described rickets resistant to vitamin D therapy, which at that time they regarded as differing from ordinary rickets only in severity and failure to respond to the usual therapy, but healing with massive doses of vitamin D.

Scorbutic changes in the spine are rarely mentioned. In 1 case observed by us a moderate degree of demineralization of the dorsal and lumbar segments occurred but no specific deformities were noted.

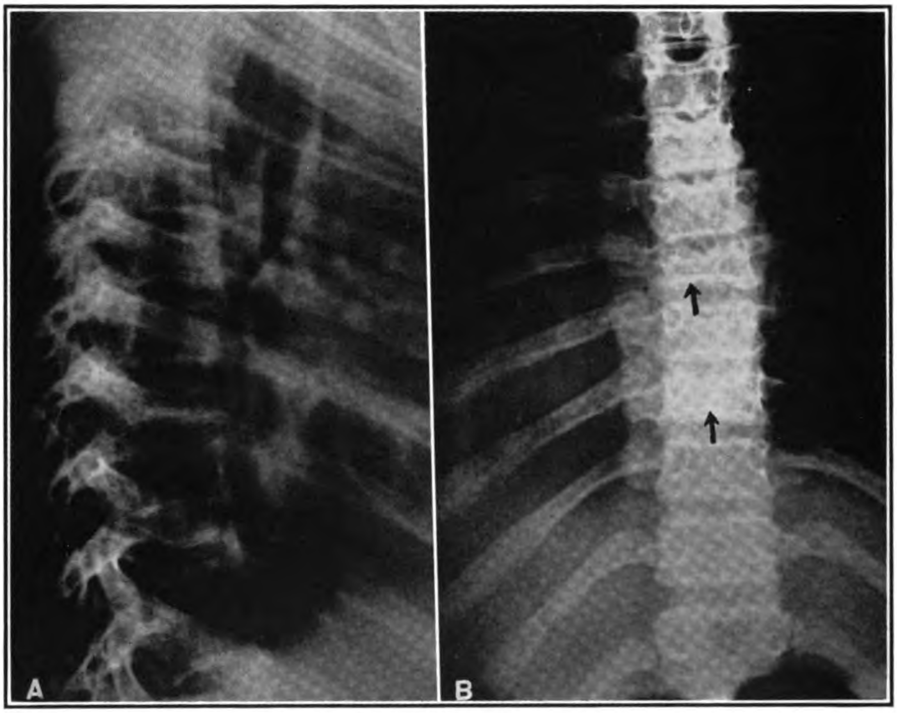

FIG. 124.—Lateral and anteroposterior spine roentgenograms in a case of rickets, with hypocalcemia. The vertebral bodies are diffusely compressed (arrows) and the intervertebral discs are bulging.

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HYPERVITAMINOSIS A

The bony changes associated with this condition usually are referable to the long bones with periosteal elevation and subperiosteal calcification involving these bones as well as the clavicles and the ribs. No spine changes have been described in the reported cases, most of which occurred in children (Caffey, 1951; Bifulco, 1953).

We had the opportunity of studying intensively a patient who had been taking over 500,000 units of vitamin A daily for over eight years. This patient, who was reported in detail by Gerber, Raab and Sobel (1954), was studied radiologically by the present author, and the findings were of great interest. Chief among these were changes in the spine which were quite similar to those usually associated with rheumatoid spondylitis A pronounced straightening of the spine was noted together with decalcification of moderate degree of the respective vertebral bodies. No change at the intervertebral spaces could be demonstrated. Of chief interest was the intense decalcification and loss of articular surfaces of the apophyseal joints of the lumbar spine. The bony margins of the sacroiliac joints, however, retained a normal bony density. Other changes which were noted were calcium deposits in the insertions of the various ligaments such as those at the anterior tibial spine, the insertion of the quadriceps tendon, the periphery of both hip joints, in the vicinity of the capsules of the shoulders, and at the iliolumbar ligaments.

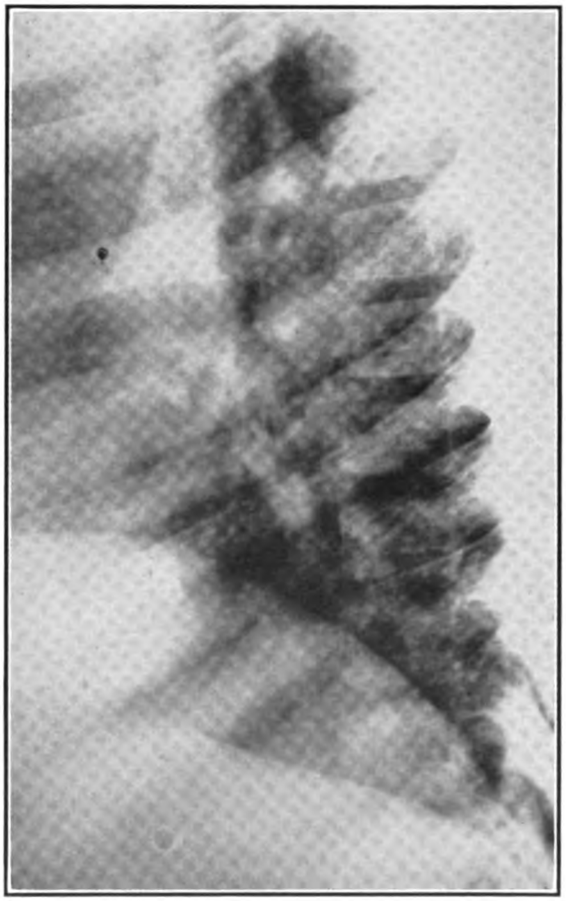

FIG. 125.—Lateral thoracic spine roentgenogram in case of vitamin D resistant rickets. Diffuse osteomalacic and granular changes in the vertebral bodies is present. The intervertebral discs are well maintained in this case.

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HYPERVITAMINOSIS D

In a report by Christensen, Liebman and Sosman (1951) no vertebral involvement in cases of hypervitaminosis D was recorded.

Recently, however, DeWind (1961) reported the case of a five and one half year old boy who had been subjected to an excessive intake of vitamin D over a long period of time. A pronounced degree of osteosclerosis followed. Radiographic examination of the spine disclosed dense square-appearing vertebral bodies. There was a generalized increase in bone density as well. The osseous changes were reversible, and were attributed to the effect of withdrawal of the vitamin D on the hypercalcemia. DeWind believed that the changes seen in his patient resembled somewhat that seen in severe cases of 'idiopathic hypercalcemia.'

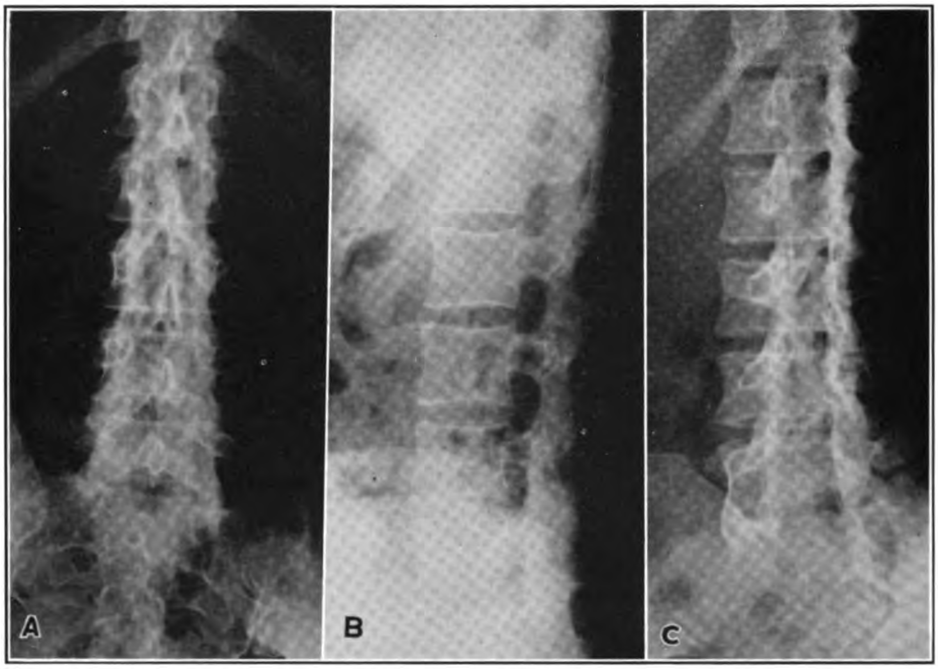

FIG. 126.—Hypervitaminosis A in an adult woman. The lumbar spine is straightened, and calcification is noted along the ligamenta flava. The articular surfaces of the lower apophyseal joints are closely apposed and roughened. The picture simulates rheumatoid arthritic changes.

REFERENCE

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IDIOPATHIC HYPERCALCEMIA OF INFANCY

This infrequent condition is attributed to an abnormal sensitivity in some children to Vitamin D, resulting in excessive absorption of calcium from the gastrointestinal tract with consequent hypercalcemia, renal calcinosis, acidosis, failure to thrive and ultimately death. Other causes which have been postulated but which do not appear tenable include primary disease of the kidneys or parathyroid glands and excessive intake of alkali.

Because of the high calcium level, marked muscular hypotonia, anorexia, vomiting, constipation and retarded growth occur. Persistent hypercalcemia results in permanent damage, with marked retardation of growth, toxic effects on the central nervous system, heart, kidneys, craniostenosis and mental retardation. Some children present a characteristic "elfin" facies, and association with cretinism has been observed.

Radiographic examination reveal extensive osteosclerosis, soft tissue calcifications and defective ossification. The osteosclerotic changes in the long bones appear at their ends, with ricket-like deformities and osteosclerotic horizontal lines. The bones appear to be rather soft, being bowed and having the epiphyseal ends invaginated into the shafts to some degree. The vertebræ partake in the osteosclerosis, their bodies particularly presenting increased density in a frame-like fashion, leaving a relatively radiolucent center. The bodies also are small.

A benign form of hypercalcemia in children is known to occur, which is not as striking and is relatively self limited. The radiologic changes are minimal. It is not known whether a relationship exists between the two forms of hypercalcemia.

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OCHRONOSIS

This is a rare and probably hereditary metabolic anomaly of unknown origin, usually appearing in adults in middle life, and which affects both sexes equally. The disease is characterized by excretion of alkapton in the urine, rendering the urine dark or brown-black after exposure to the air. The chemical substance excreted has been identified as homogentisic acid, which is an intermediary product in the metabolism of phenylalanine, beyond which that amino-acid and tyrosin are not metabolized. A few cases have been discovered in infants because of brownish or black discoloration of the diapers. The condition is usually asymptomatic until middle age, at which time the patients become poorly nourished and appear chronically ill. Their skin becomes a mottled yellowish or brownish color, particularly over the head, neck and torso and to a lesser degree over the extremities. The ear lobes appear nodular and rigid and become a discolored greyish blue color. The nose also may acquire a bluish tint due to the deposit of homogentisic acid, and the scleræ likewise assume a slate blue or purplish appearance, with triangular brownish patches. Clubbing of the fingers occurs and discoloration of the hands, the axillæ and the genital regions because of pigment deposits occur. These people usually develop a

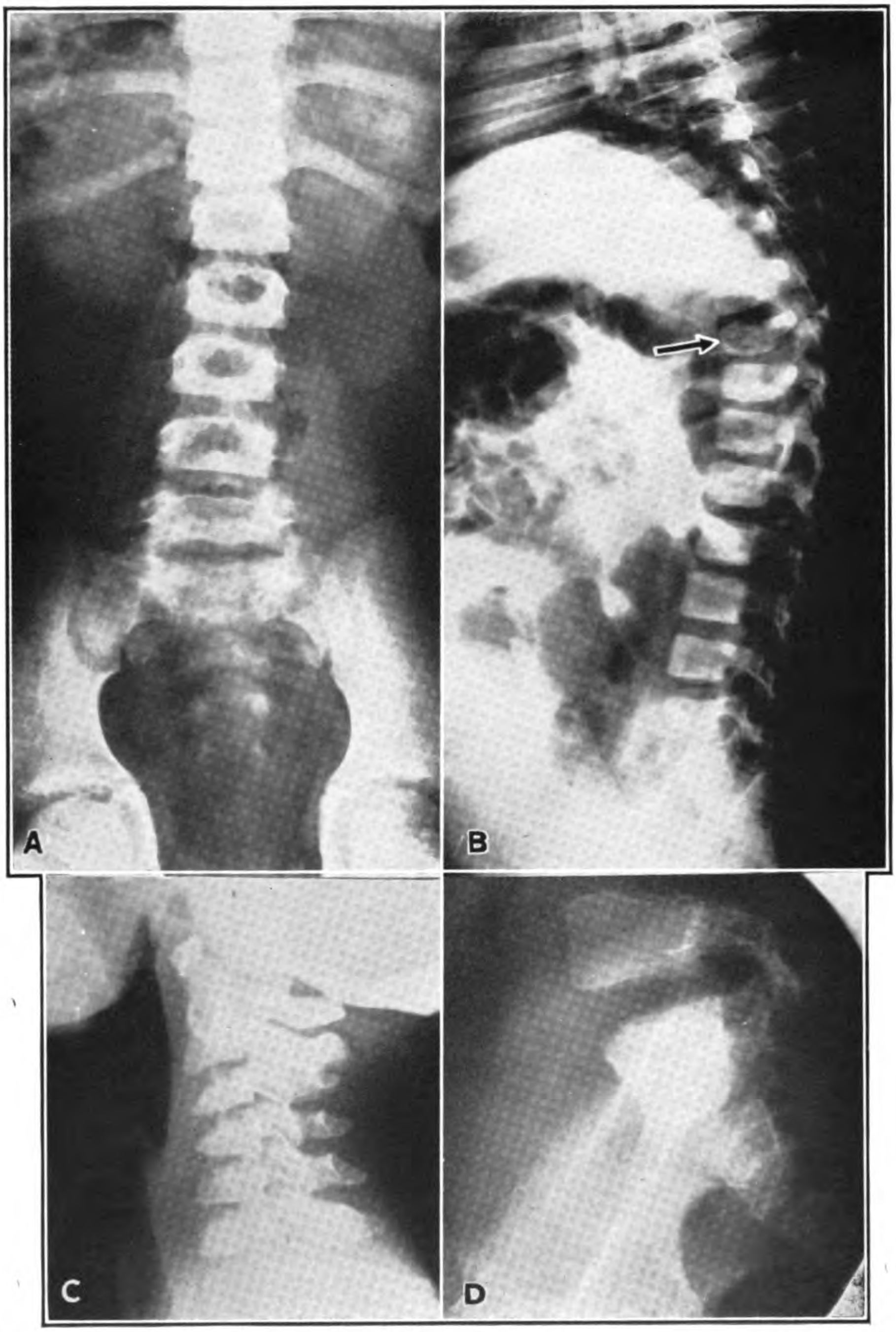

FIG. 127.—*A* and *B*, Anteroposterior and lateral thoracolumbar spine in case of lipodystrophy. There is a dorsal curvature at the thoracolumbar junction. The body of the tenth dorsal vertebra at the apex of the curve is small (arrow). *C*, Same case, lateral cervical spine. Note the anterior wedging of the bodies of the third and fourth cervical vertebræ. *D*, Same case. Lumbosacral articulation. The fifth lumbar vertebral body is wider anteriorly than posteriorly, and its bony density is slightly diminished. The cancellous markings are hazy.

dorsal kyphotic curve with loss of the lumbar curve. Radiologic examinations of the spine show a hypertrophic ankylosing process with osteoporosis of the vertebral bodies. The dorsal kyphosis may be prominent. Vertical or linear striations may be seen in the vertebral bodies, which may be reduced in height. Most striking is a diffuse calcification of practically all the intervertebral discs, which is considered diagnostic of this condition.

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Progressive Lipodystrophy

This condition is characterized by a loss of the normal fatty tissue in the upper half of the body. The adiposity of the lower trunk, pelvic regions and lower extremities becomes increased so that the lower half of the body is disproportionately bulky. The disease occurs more often in females, and may occur from childhood well into adult years. As a rule the patients are able to continue their activities, but may complain of easy fatiguability. The loss of fat may be progressive. According to Ziegler (1928) the hypothetical causes are neurotrophic and endocrine disturbances, alteration of fat metabolism, race, heredity, infection and congenital defect. No one of these has been definitely identified as a single etiologic factor. The disease is neither hereditary nor familial.

No skeletal alterations have been mentioned as part of the picture of lipodystrophy. One patient, a twelve-year-old boy, seen by us showed changes in the spine (Fig. 127). The anterior aspects of the middle cervical vertebræ were sharpened and angular. The thoracic vertebræ were not disturbed, but a definite convexity of the thoracolumbar junction was present.

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Chapter

4

Inflammatory, Degenerative and Noxious Diseases of the Spine

ANKYLOSING SPONDYLITIS (RHEUMATOID SPONDYLITIS, MARIE-STRÜMPELL DISEASE, ANKYLOSING SPONDYLARTHRITIS), AND RHEUMATOID ARTHRITIS.

Ankylosing spondylitis is a fairly common disease of still unknown etiology which usually appears in males during the second to fourth decades, and is not uncommon even later. It has a marked inflammatory component, occurs in siblings and is likely to affect the offspring of similarly afflicted parents. Mental shock, trauma and acute infectious disease sometimes precipitate its onset or aggravates its manifestations. The progress of the disease may be slow or rapid, with remissions and exacerbations. The advent of steroid therapy is a noteworthy step in the management of this disabling condition, but the prognosis remains rather gloomy insofar as progressive disability is concerned. The relief of symptoms with *x*-ray therapy has made a place for this modality, but the possibilities of leukemogenic effect is a deterrent to its usage.

The question whether ankylosing spondylitis and rheumatoid arthritis are the same basic disease is still largely unresolved. The striking resemblance of the histopathologic changes in the afflicted joints is noteworthy, as is the presence of occasional peripheral joint involvement in patients with ankylosing spondylitis and vertebral column disease in individuals whose peripheral joints are mainly involved. In Great Britain ankylosing spondylitis is separated from rheumatoid arthritis on the basis of differences in the pattern of joint involvement, the difference of incidence in males and females, the absence of streptococcal and sheep red cell agglutinins in ankylosing spondylitis, the absence of subcutaneous nodules in ankylosing spondylitis and some histologic features. Gold therapy has been found more effective in rheumatoid arthritis, while *x*-ray therapy is more beneficial in ankylosing spondylitis. Another point is the existence of severe osteoporotic changes in the affected peripheral joints in patients with rheumatoid arthritis contrasted with its relative absence in the occasional case of ankylosing spondylitis with peripheral joint involvement. It appears that the main differential points are those referring to the involved areas, the sex incidence and the pattern of bone change in the peripheral joints. None are absolute, and the conclusion must be reached that we still cannot make more than a clinical differentiation, realizing that certain aspects of both diseases are present in either.

Low back pain is one of the leading initial complaints of patients with ankylosing spondylitis, sometimes associated with changing sciatic radicular symptoms. Later the pain involves the lumbosacral and thoracolumbar areas. Initial involvement of peripheral joints sometimes confuses separation of the entities, particularly if the articular symptoms are migratory and of relatively short duration. During the active stages fever, lassitude and weight loss accompanies the stiffness of the back, and the kyphotic stance may appear rather rapidly. The period of activity may be relatively short or relentlessly progressive over a period of several years, producing a fixed, rounded configuration of the thoracic spine and consequent forward thrust of the neck. Spinal involvement with rheumatoid arthritis usually is in the cervical region, and may appear in relatively young children with ankylosis of the posterior cervical articulations. The peripheral joints are frequently involved in rheumatoid arthritis, while such change is relatively uncommon with ankylosing spondylitis. Sacroiliac involvement in rheumatoid arthritis is not present, while it is frequent and early in ankylosing spondylitis. Another clinical point is that ankylosing spondylitis is rare before puberty, while rheumatoid arthritis may appear even in children.

The earliest pathologic change associated with rheumatoid disease is marked proliferation of the synovial tissues, the articular capsules and surrounding soft tissues of the joints. Vascular granulation tissue then appears over the articular cartilages. Concomitantly, a similar inflammatory process takes place in the connective tissue elements in the bone below the articular cartilage. The inflammation then extends through the zone of temporary provisional calcification and destroys the articular cartilage nearest the bone. At the same time the articular cartilage itself is attacked. These two layers of active granulation tissue have marked potentialities for forming fibrous and bony ankylosis. This replacement of normal synovial tissue by granulation tissue, eroding cartilage and invading bone, results in marked deformities of the joints with loss of function and ankylosis.

In the early stages of the disease, increased synovial fluid of an inflammatory nature appears in the involved joints. With recession of the attack the joint effusion disappears, but in the chronic stages the hyperemia and inflamed synovial membranes become prominent. At first the synovial membrane is engorged and swollen, but retains its smoothness. Later its surfaces become granular and attached firmly to the articular cartilages as a sheet of thickened vascular fibrous tissue termed pannus. Ankylosis of the joints may occur when a pannus firmly binds two opposing surfaces, or when the surfaces meet because of ulceration through their cartilaginous boundaries leading to apposition of bony structures (Hare, 1940; Steinberg, 1942; Oppenheimer, 1945). Gibson (1957) reported that biopsies from the sacroiliac and manubriosternal joints indicated change from fibrocartilage to fibrous tissue to chondroid and later to bone, with no truly inflammatory phase. Serial biopsies from the tips of the spinous processes showed inflammatory destruction followed by excessive bone formation. In the posterior spinal joints adhesions form rapidly after a villous synovitis, followed by chondroid metaplasia, ossification and ankylosis. In the intervertebral discs ossification occurs in the peripheral fibers of the annulus fibrosus and spreads from the margins of the epiphyseal rings of the body on either side to meet in the midline, producing first small spur formations and later a bamboo spine.

Rheumatoid disease is not entirely a disease of the articular structures, but as a disease of connective tissue in general, may produce changes in the cardiovascular system, the soft tissues of the skin in the form of rheumatic nodules, and the peripheral nerves with a somewhat similar nodular change. In the later stages the bony structures are involved to a considerable degree with atrophy and erosions in localized areas. These changes are caused by extension of vascular granulation tissue from the joint through the cartilage into the adjacent bone.

According to Forestier (1939) the sacroiliac joints are involved earliest, changes having been observed by him in over 98 per cent of 153 cases of ankylosing spondylarthritis. The involvement of these joints may be bilateral or unilateral. Overgaard (1945) mentioned three radiologic stages of involvement of the sacroiliac joints. The first was scattered, slightly cloudy opacities in the para-articular bone, and slight spotted decalcifications of the subchondral osseous tissue with vague and blurred demarcation. The second consisted of bony bridges which tended to obliterate the articular spaces. The third stage was that of complete ankylosis. There is apparently uniformity of opinion that the earliest change associated with rheumatoid spondylitis is a decalcification of the bone adjacent to the sacroiliac joints producing blurred and indistinct margins.

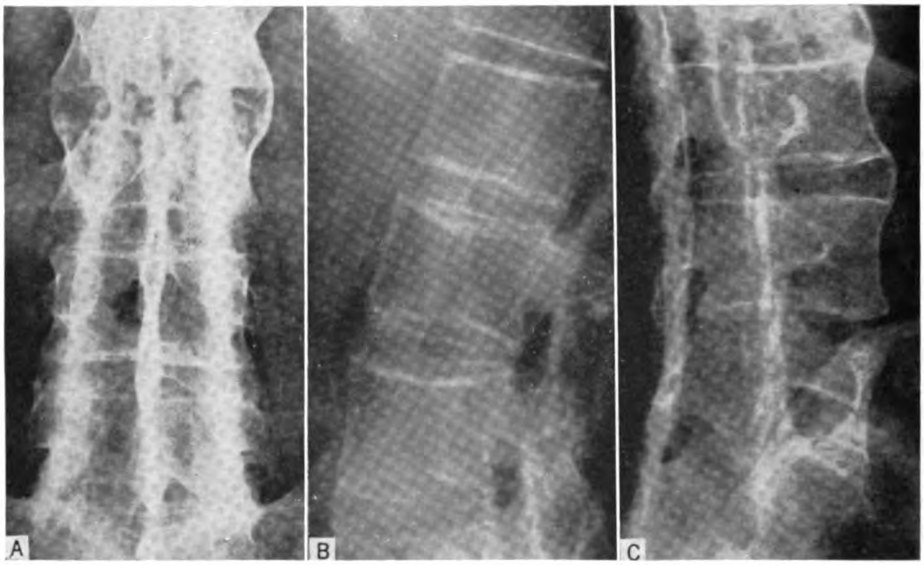

FIG. 128.—Rheumatoid spondylitis in a 36-year-old male.

A, Anteroposterior view. Calcification of the paraspinal ligaments is present in the upper lumbar region. The supraspinal ligament uniting the apices of the spinous processes is calcified, and the posterior intervertebral joints present as heavy, linear markings to either side of the midline because of ankylosis. *B*, The vertebral bodies are osteoporotic. Some calcification is present in the anterior spinal ligament. The intervertebral discs are slightly widened. The margins of the vertebral bodies appear sclerotic. *C*, Oblique roentgenogram. The thin calcification of the paraspinal ligament is better visualized. The apophyseal joints are narrowed and their margins are sclerotic.

The early radiologic spinal changes of ankylosing spondylitis are manifested also in the apophyseal joints. These first present merely a narrowing and loss of density of opposing articular facets. Later this may progress to an actual rarefaction of bone and irregularity of the articular margins. Progressively narrowed joint spaces and ankylosis eventually results. These changes in the apophyseal joints are quite difficult to interpret, particularly in the early stages of the disease. Caution is urged in evaluating minimal changes in the apophyseal joints. As the disease progresses, demineralization of the vertebral bodies occurs together with calcification in the anterior and lateral spinal ligaments.

The appearance of the intervertebral discs in patients with ankylosing

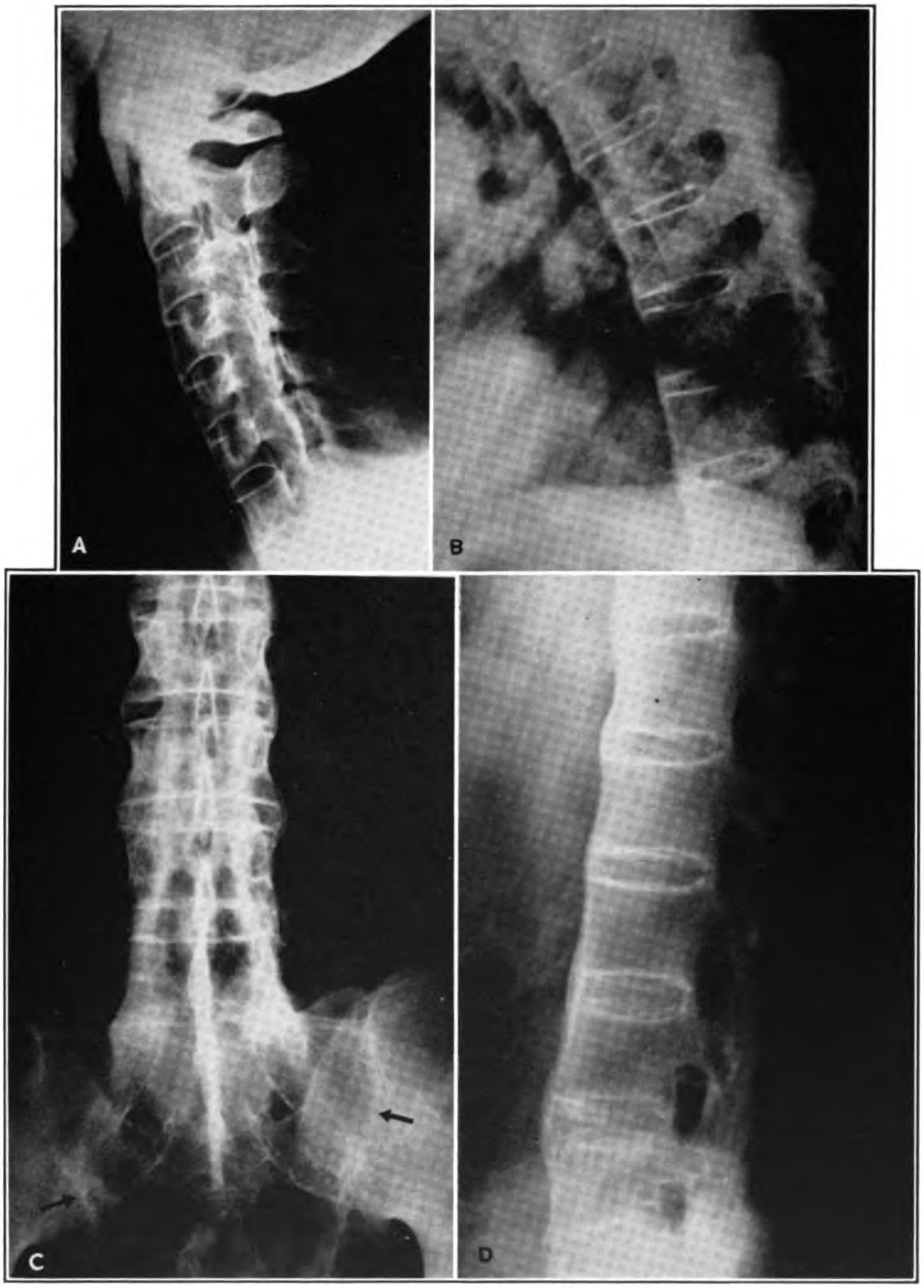

FIG. 129.—*A*, Lateral neck roentgenogram, ankylosing spondylitis. Note the biconvex appearance of the intervertebral discs, the vertebral osteoporosis, calcification of anterior spinal ligament and the straightened cervical lordotic curve. *B*, Same case. Lateral dorsal spine with calcification of anterior spinal ligament and similar changes to those in the cervical spine. *C*, Same case, anteroposterior view of lumbosacral spine. Calcification of lateral spinal ligaments and ligamenta flava. Note the loss of the articular markings of the sacroiliac joints, with replacement by a thin linear fused joint line of increased osseous density (arrows). *D*, Same case. Lateral lumbar spine. The anterior spinal ligaments are densely calcified. The spine is straight. The vertebral bodies are well formed but are osteoporotic. The intervertebral spaces are quite wide. Some ossification is noted at the anterior aspect of the fourth lumbar intervertebral disc.

spondylitis has evoked discussion. For some time it was believed that the intervertebral discs retained their normal thickness throughout the disease. This was mentioned in 1946 by Stecher and Hauser, and in 1951 by Solovay and Gardner. Romanus and Ydén (1952) described narrowing of the intervertebral discs in about one-third of their patients with ankylosing spondylitis. Pursuing this line of investigation with discography, they found that pathologic changes in the intervertebral discs were not uncommon and described the association of herniation of the nucleus pulposus with abnormal disc lesions.

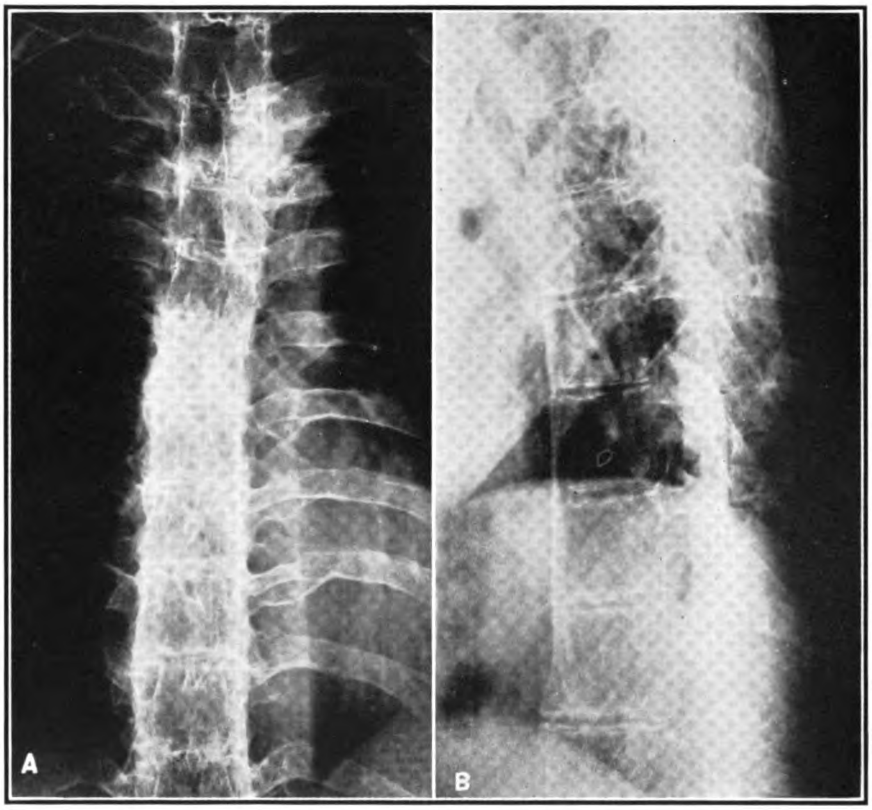

FIG. 130.—Anteroposterior and lateral roentgenograms of thoracic spine in rheumatoid spondylitis. The spine presents a straightened "poker spine" configuration. Note the pronounced osteoporosis, the narrowed intervertebral discs and the calcified anterior and lateral spinal ligaments. The apophyseal joints are fused and sclerotic, forming parallel streaks. The costovertebral joints also are ankylosed.

A review of our material disclosed that the appearance of the intervertebral discs is quite variable in patients with ankylosing spondylitis. In those individuals who had the disease during the early years of life the spine became fixed in a relatively straight position, and in these cases the intervertebral discs as a rule maintained themselves fairly well. In some instances the discs became rather wide, particularly in the lumbar spine. However, in the dorsal spine slight, and sometimes considerable narrowing of the intervertebral discs was noted. This change was apparent in individuals who had developed a pronounced degree of calcification of the para-

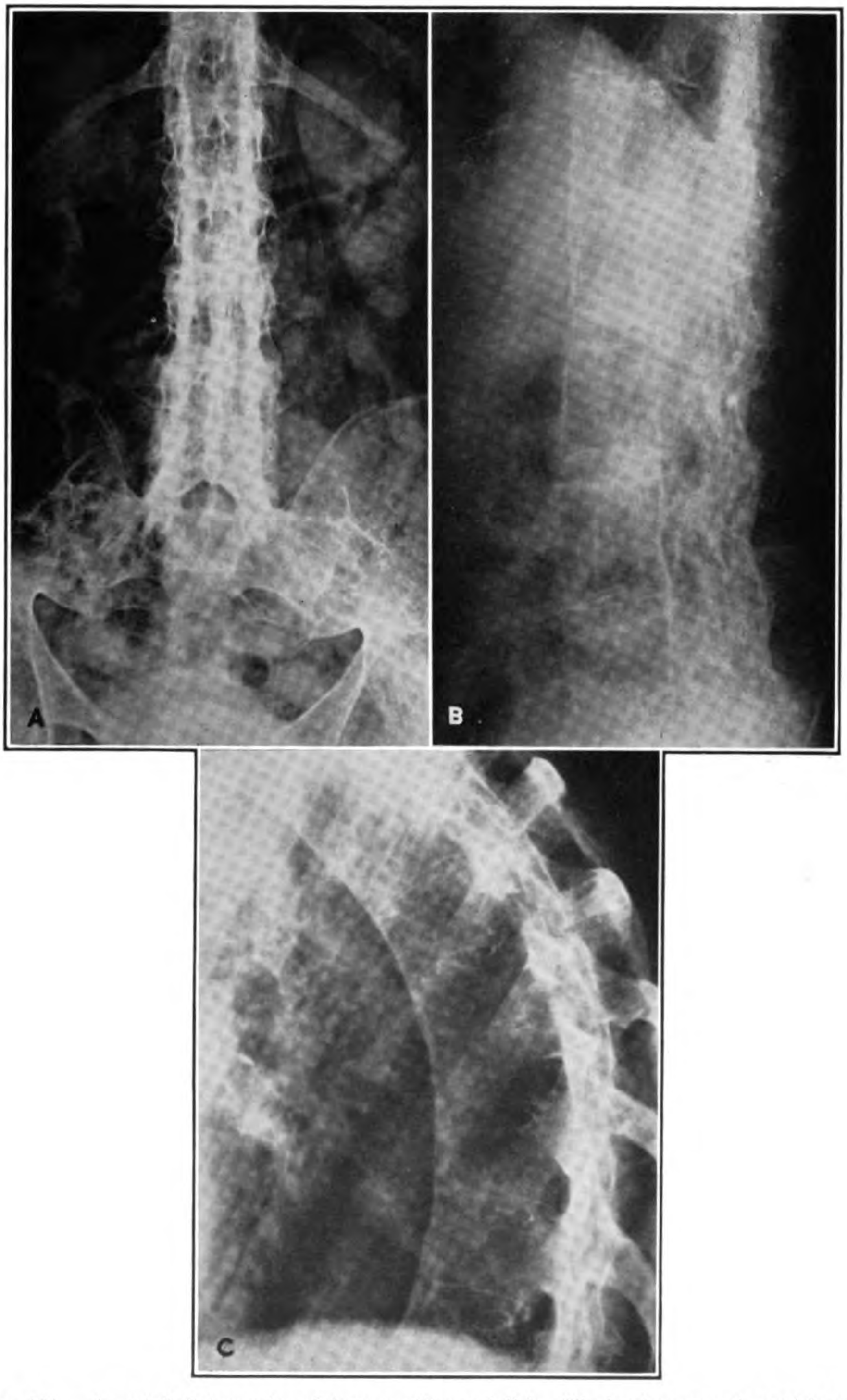

FIG. 131.—*A*, and *B*, Poker-spine of rheumatoid spondylitis. The lumbar curve is straightened. The vertebræ are markedly osteoporotic, and the intervertebral discs are slightly narrowed. The calcification of the paraspinal ligaments is mild. The apophyseal joints are fused, and the ligaments between these and the supraspinal ligament are densely sclerotic. *C*, Another patient with rheumatoid spondylitis with a pronounced dorsum rotundum. The spine is fixed. The vertebral bodies are markedly demineralized. The anterior spinal ligament is calcified. The margins of the intervertebral discs are barely perceptible.

spinal ligaments leading to the so-called "poker spine." In some cases the narrowing of the disc was so marked that the margins were practically in apposition one to another. It was not uncommon to note areas of linear increased density at the superior and inferior margins of some of the vertebræ which had closely approximated each other. In other patients in whom the disease was less precipitous in its advance, and in whom the vertebral bodies retained a good degree of osseous density, changes in the intervertebral discs seemed to parallel those usually associated with that particular age group. In individuals in early middle life, in whom a moderate dorsum rotundum had appeared, hypertrophic changes at the articular margins of the discs were not uncommon. In these patients, narrowing of the intervertebral spaces, particularly along the middle of the arc of the dorsum rotundum, was the rule rather than the exception. These changes seemed to be most pronounced in the upper two-thirds of the thoracic spine. The lower third of the thoracic spine maintained the usual height of the intervertebral discs for a longer time. The discal changes probably bear some specific relation to ankylosing spondylitis even though there is no synovial membrane in the intervertebral fibrocartilaginous joints.

When demineralization of the vertebral bodies occurs, the intervertebral discs assume a somewhat biconvex appearance. While small Schmorl's nodes are occasionally noted in the lower dorsal and upper lumbar region, their appearance in these regions, and more particularly their appearance in the remainder of the dorsal and lumbar spine, is uncommon.

In those in whom the progress of the spondylitis was more insidious, normal osseous density appeared to be retained for a longer period of time, even though considerable calcification appeared in the spinal ligaments. These individuals also had a tendency to develop a moderate dorsum rotundum.

When reviewing films of the dorsal spine it may be of interest and diagnostic help to observe the appearance of the angle of Louis. With ankylosing spondylitis it is not infrequent to note articular and bony changes in the manubriosternal joint. These may appear as widening of the joint space with sclerotic and erosive bone edges (Solovay and Gardner, 1951). In two recent cases of young men believed to have early manifestations of ankylosing spondylitis I have observed similar changes in the sternoclavicular joints, best delineated on body section roentgenograms. A similar appearance of the ischial tuberosities and the symphysis pubis attributed to subcortical bone destruction is mentioned by Middlemiss (1956).

As a rule, the usual lumbar lordotic curve was either straightened or shallower than normal, even though considerable dorsum rotundum had appeared. Paravertebral calcification appeared to be most intense in the vicinity of the lumbar and thoracic spine but similar changes in the cervical spine were by no means uncommon. The appearance of the paraspinal calcification was not uniform. In several instances calcification was quite pronounced in the lumbothoracic region and absent in the upper dorsal and lower lumbar areas. When the disease had been present for a considerable time, changes in the apophyseal joints were readily apparent. The sacroiliac joints showed almost complete loss of their bony margins, the joint spaces being seen as thin white lines where the opposing surfaces of the sacrum and iliac bones met.

The effect of *x*-ray therapy on ankylosing spondylitis is beneficial. Favorable results have been reported by many observers, particularly in the early stages of the disease (Smyth, Freyberg, and Lampe, 1941); Hemphill and Reeves, 1945; Borak and Taylor, 1945; Baker, Conrad, Reeves and Hoyt, 1950). In some cases of a mild or moderate nature the process can be arrested or sometimes completely resolved, with improvement in the roentgenographic appearance of the involved spine. However, such physical changes cannot be anticipated if there has been actual destruction of cartilage and bone surfaces. The beneficial effects of cortisone and corticotropin are well known and require no discussion here. However, these efficient agents bear with them certain risks incident to their demineralizing effects. A report of pathologic fractures developing in 5 female patients

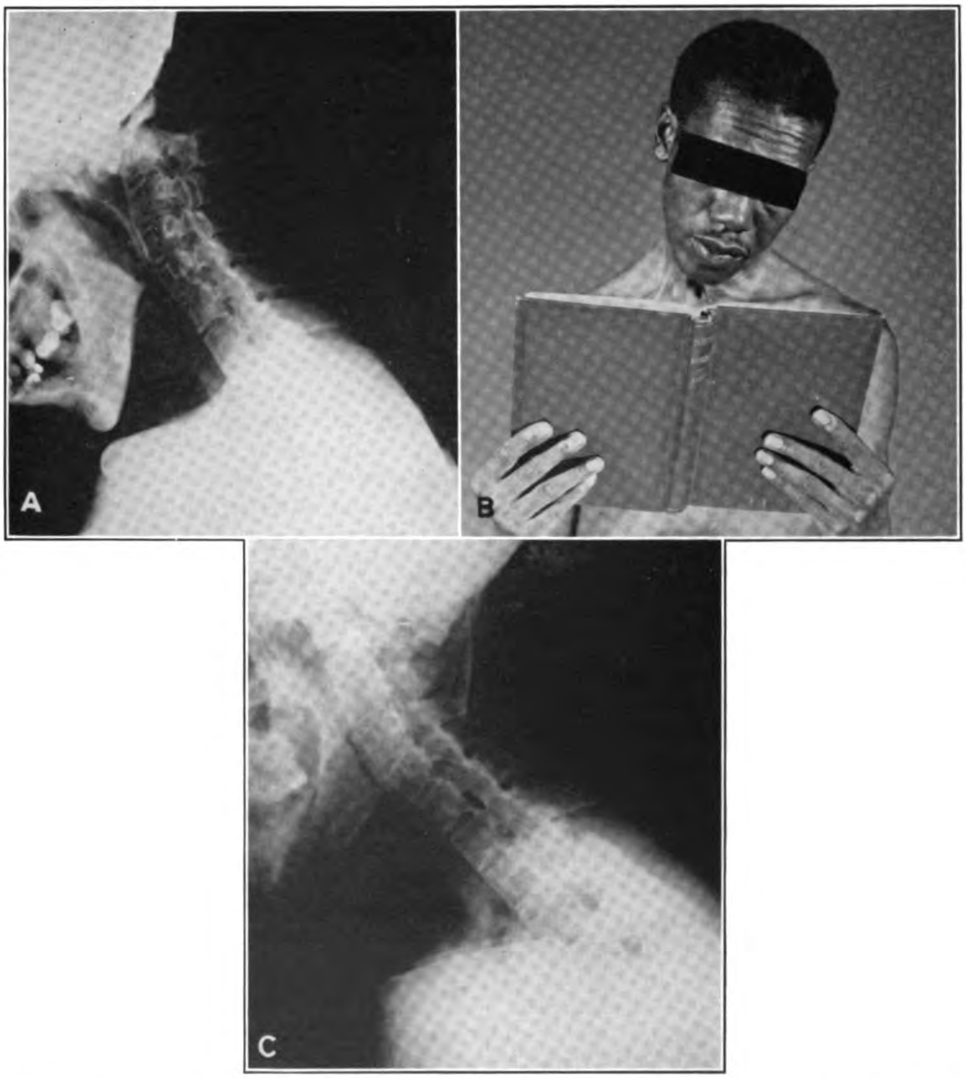

FIG. 132.—*A*, Lateral spine roentgenogram showing anterior dislocation of the first on the second cervical vertebræ incidental to rheumatoid spondylitis. *B*, Note position of head of this patient. *C*, Same case, post-reduction lateral cervical spine roentgenogram. (Courtesy of Dr. M. E. Margulies, Veterans Administration Hospital, Brooklyn, N. Y.)

with rheumatoid arthritis treated with cortisone over a long period of time was reported by Demartini, Grokoest and Ragen (1952).

An interesting complication occasionally observed in patients with advanced ankylosing spondylarthritis is reduced efficiency of the costovertebral mechanisms secondary to ankylosis of the joints between the ribs and the dorsal vertebræ. In such instances the effect of the costovertebral immobility is to reduce the efficiency of the respiratory and cough mechanism, resulting in recurrent attacks of pulmonary disease (Hamilton, 1949).

The clinical picture of ankylosing spondylitis has also been associated with a number of other conditions totally unrelated to the basic concept of the disease. Examples of these are brucellosis, which has been reported as producing changes in the sacroiliac joints practically indistinguishable from those associated with rheumatoid arthritis (Steinberg, 1948), intestinal amebiasis (Rappaport, Rossien and Rosenblum, 1951), and that form of arthritis which not infrequently is observed in women of menopausal age (Greenblatt and Kupperman, 1946). Abel (1950) reported that patients with paraplegia may present sacroiliac joint changes similar to those seen with ankylosing spondylitis. Acute or chronic pelvic visceral sepsis may be responsible for destructive lesions of the sacroiliac joints.

Destructive bone lesions with ankylosing spondylitis are rare. Of great interest is the case reported by Baggenstoss, Bickel and Ward (1952) in which granulomatous nodules occurred in the body of a vertebra, producing destructive changes. *X*-ray examination disclosed destruction and wedging of the body of the twelfth dorsal vertebra. At necropsy there was destruction of this vertebra and a nodule in the third lumbar vertebra. These had been considered myelomatous until histologic investigation revealed bony changes including palisading of fibroblasts around a central necrotic area and destruction of adjacent bone trabeculæ similar in nature to lesions present in the heart and pericardium. The necrotic focus was of a granulomatous inflammatory nature such as occurs in the viscera and subcutaneous tissues. They also mentioned 3 other cases with destruction of vertebræ without histologic confirmation. Wholey, Pugh and Bickel (1960) reported ten cases with localized bone destruction on either side of the intervertebral disc. This was more pronounced anteriorly and was followed by spontaneous fusion and ankylosis. A certain similarity to the changes present with tuberculosis, osteomyelitis and other granulomatous lesions was observed. When fairly extensive bone destruction was present metastatic disease had to be considered and biopsy was required sometimes to make the diagnosis. The bone destruction is much less conspicuous than the other more obvious effects observed with rheumatoid spondylitis. In two cases biopsies revealed histologic changes compatible with rheumatoid inflammation. Gibson (1957) illustrated a case of rheumatoid arthritis in which the cervical spine of an adult male patient presented destruction of the intervertebral disc between the third and fourth cervical vertebræ leading to subluxation with cord pressure.

Atlanto-axial dislocation occurs both in rheumatoid arthritis and ankylosing spondylitis. There may be pain in the cervical region together with neurological deficits which vary with the extent of the spinal cord pressure resulting from the forward displacement of the atlas on the axis. The earliest radiologic change is a widening of the space between the anterior aspect of the dens and the posterior aspect of the anterior arch of the atlas. Normally this space measures about 2.5 mm. Sharp and Purser (1961) regard a separation of more than 3 mm. as abnormal after the age of 44 years, although a gap of 4 mm. may be normal in younger individuals. With more advanced displacements a lateral shift and some rotational defects occur, producing torticollis and a forward position of the head together with a flattening of the cervicooccipital curve. The early changes are best identified on flexion films. In two cases noted by us there also appeared to be a definite degree of demineralization of the second cervical vertebra, particularly of the dens. Atlanto-axial dislocations are observed less frequently in patients with ankylosing spondylitis, but in these patients is more likely to be accompanied by symptoms. It is important to keep in mind that tetraplegia and death may follow sudden compression of the medulla or upper cervical spinal cord, and these patients should be handled with great care. It is advisable not to attempt flexion examinations unless indicated by the inability to make the diagnosis from a resting lateral cervical spine roentgenogram. In patients with an obvious atlanto-axial dislocation little is to be gained by exaggerating the defect by making the patient flex the neck. However, when there are complaints of pain together with no definite displacement of the dens, flexion examinations are important in determining the amount of movement the dens can undergo. The demonstration of widening of the space between the dens and anterior arch of the atlas then becomes important.

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JUVENILE RHEUMATOID ARTHRITIS

Juvenile rheumatoid arthritis is often referred to as Still's disease and its pathologic manifestations are much the same as those of rheumatoid arthritis. In addition, splenomegaly, lymphadenopathy and occasionally hepatomegaly are seen fairly often. The joints most commonly affected in the polyarticular form in children are those of the hands and wrist, the feet, ankles, knees and tempero-mandibular joints. Accelerated epiphyseal growth causes elongated configurations of the long bones (Coss, 1946). The cervical spine is involved in about 13 per cent of the cases. The disease occurs more often in females and is seen most frequently in the second to third years of life. The children present a delicate, almost bird-like fragile appearance.

In early cases of juvenile rheumatoid arthritis no radiologic changes may be apparent in the spine. Later, the *x*-ray findings are those of spondylitis with relatively little calcification of the paraspinal ligaments. Some degree of decalcification of the vertebral bodies may take place early in the disease, but this is a very difficult alteration to evaluate. The lumbar lordotic curve may become straightened. As with the adult form of rheumatoid spondylitis thinning and approximation of the apophyseal joints are of considerable diagnostic importance. The intervertebral discs, according to Buckley (1943), do not become thin but may even expand and complete ankylosis may eventually result. Saenger (1950), in a report of 4 cases in children, the oldest of which was fourteen years of age, described a syndrome in which narrowing of the intervertebral discs appeared about two to four weeks after the onset of clinical symptoms. In these individuals pain was referable chiefly to the hip. Some demineralization or destruction of parts of the vertebræ appeared. Narrowing of the intervertebral spaces persisted for about four to twelve weeks, when sclerotic changes began to be evident in the vertebræ. During the next two to eight months there was gradual widening of the intervertebral spaces with sclerosis and new bone formation in the area of destruction of the opposing bone margins. In very young children restitution to normal occurred within a year.

Later in life ankylosis of the apophyseal joints of the upper cervical vertebræ may occur. These vertebræ tend to assume a diminished antero-posterior diameter as well as height. Their intervertebral discs may narrow or disappear, resulting essentially in a small, fused upper cervical vertebral column.

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REITER'S SNYDROME

This is a disease of unknown etiology which in its complete form, consists of a triad of conjunctivitis, urethritis and arthritis. The disease is quite infrequent and has been attributed to a number of infectious origins, none of which is considered conclusive. Biopsy of synovial membrane in 1 case was reported by Bauer and Engelman (1942) as differing from the changes associated with either infectious or rheumatoid arthritis. As a rule, the joints most frequently involved are those of the elbows and wrists and, to a greater extent, the knees and ankles. Involvement of the spine and sacroiliac joints is relatively infrequent. In the 25 cases reported by Hollander, Fogarty, Abrams and Kydd (1945), mild and nonspecific spinal involvement was encountered in only 2 cases. The characteristic roentgenographic abnormalities were those of a diffuse osteoporosis which disappeared as the condition cleared. Hollander and his co-workers studied synovial membrane removed from the right knee joint of 1 of their patients. In the course of removing the material it was noted that the cartilaginous surfaces of the joint were undisturbed. Their observations confirmed those of Bauer and Engelman (1942), in that an intense hyperemia and cellular reaction limited to the superficial layers of the synovium was found. They considered the disease process as distinctly different from that seen with rheumatoid arthritis.

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WERNER'S SYNDROME

According to Thannhauser (1945) the distinguishing characteristics of this hereditary familial condition includes a premature graying of the hair and baldness, shortness of stature, scleropoikiloderma, trophic ulcers of the legs, juvenile cataracts, hypogonadism, a tendency to diabetes, osteoporosis, calcification of the blood vessels and metastatic calcification. It has been referred to as progeria of the adult. The radiologic changes, insofar as the spine were concerned, were those of an advanced spondylitis inconsistent with the patient's age. Another example of this condition was reported by Pomeranz (1948).

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PSORIATIC ARTHRITIS

This condition is considered by some to be a variant of rheumatoid arthritis associated with psoriasis. It has been noted that the clinical activity of the disease varies with the extent and activity of the psoriatic condition. In a case described by Jungmann and Stern (1944) fusion of certain vertebræ was observed, together with advanced changes involving the articulations of the extremities. In another case reported by Nunemaker and Hartman (1950) no spine changes were observed. Sherman (1952) described atrophic changes in the hands. In 2 cases observed by us the changes were most marked in the hands and wrists, with no definite alteration in the structure of the vertebral column.

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TUBERCULOSIS OF THE SPINE (POTT'S DISEASE)

Tuberculous osteomyelitis of the vertebral column is caused by hematogenous extension of the tubercle bacilli into the medullary spaces. With involvement of a single vertebra, Key (1940) designated the types of involvement as central, epiphyseal, or anterior, depending upon the initial site of infection. This occurs most commonly in the vicinity of the upper or lower vertebral cartilaginous plates where the blood supply is ample. The caseating effects of tuberculosis produce a localized destruction of bone which slowly increases in size. Liquefaction may follow, accompanied by abscess formation. Extension of the abscess by perforation of bone into the adjacent soft tissues is along the course of the anterior or lateral spinal ligaments. Later, further extension of the abscess into the adjacent soft tissues takes place. Abscesses forming above the diaphragm tend to remain within the chest cavity or to point posteriorly, while those occurring below the diaphragm tend to enter the pelvis along the course of the psoas muscle and may point in the groin or the thigh. Less frequently abscesses burrow posteriorly and appear in the lumbar region. With further caseation, collapse of the vertebral bodies follows, accompanied by disruption of the cartilaginous plates and spreading of the tuberculous infection into the intervertebral discs. Considerable deformity of the vertebral bodies may develop, varying from an almost imperceptible irregular demineralization of the intervertebral surfaces in the early phase to a practically complete collapse of the body in advanced cases. Wedging of the involved vertebral bodies due to progressive softening is not uncommon. The end result may be an angular kyphosis which in extreme instances reaches a 90-degree angle. While the extension of a tuberculous process from the one vertebral body to another by direct continuity is by far the most frequent form of extension of spinal tuberculosis, involvement of multiple vertebral bodies distant from one another is not uncommon.

The incidence of spinal tuberculosis was reported by Bosworth and Levine (1949) to be 3.2 per cent of a total of 12,835 tuberculous patients treated at the Sea View Hospital in New York City during the period between 1940 and 1945 inclusive. This was lower than usually reported, but the lowered incidence was believed related to improved treatment shortening the period of possible metastasis. More recently Galland (1952) pointed out that the most common cause of death, tuberculous meningitis, has been practically eliminated by modern treatment. The advent of modern therapeutic agents in tuberculosis may well be expected to further reduce the incidence of tuberculosis of the spinal column.

The presence of mottled, sclerotic changes in tuberculosis of the spine is more common than is generally believed. Cleveland and Bosworth (1942) considered sclerotic changes in the involved bone to be a vascular phenomenon produced by loss of blood supply from thrombosis, endarteritis, occlusion or destruction of blood vessels by the presence of dissecting abscesses. Sclerotic changes or mottling of an involved vertebra in a known case of tuberculosis might lead to an earlier diagnosis. Auerbach and Stemmerman (1944) pointed out that in vertebræ involved in sclerotic changes, microscopic study revealed the trabeculæ to remain intact, thereby explaining the lack of collapse or other deformity in such individuals. The sclerotic changes are caused by granulation tissue and caseous material filling the marrow spaces rather than by any gross increase in the density of the involved bone. Both a caseating and productive sclerotic form of tuberculosis may coexist. In their series of 128 cases, Auerbach and Stemmerman found that 62 per cent were essentially exudative in nature, 12 per cent were essentially productive and 26 per cent showed both forms of the disease.

While it is usually considered that the repairative formation of bone is more frequent in pyogenic osteomyelitis, such changes also have been described with tuberculous spondylitis. Bridging of the vertebral bodies is regarded either as a reaction to irritation, with periosteal new bone formation, or ossification in ligamentous tissue rather than evidence of a healing process. In some cases this is extensive enough to fuse the vertebræ as effectively as a posterior surgical fusion. Guri (1947) pointed out that in a few cases healing occurred with bony fusion of the anterior aspects of the involved vertebræ. This usually followed contact between two bodies resulting from destruction of the intervertebral cartilage, or by recalcification and union of the remnants of several vertebræ which had been more extensively destroyed. The amount of concurrent deformity of the spine, the shape, structure and speed of formation of these synostoses were not uniform. The amount of protective muscle spasm may influence the final configuration of the spine.

Tuberculosis of the spine occurs more frequently in children in the upper thoracic region than elsewhere in the body. The lower dorsal and lumbar regions are more frequently affected in adults. Involvement of the cervical spine is less common. Paravertebral abscess formation occurs in about half of the adult group, and paravertebral swelling is an important early *x*-ray sign of tuberculous spondylitis. Bosworth (1945) stated that a paravertebral abscess in the thoracic region was the earliest criterion for the diagnosis of spinal tuberculosis, antedating bony or intervertebral disc changes. He reported a case in which later necropsy showed no evidence of tuberculosis grossly or microscopically, but in which films made five years before had disclosed a paravertebral shadow from the third to twelfth thoracic vertebræ with slight narrowing of the seventh thoracic disc.

Less frequent sites of involvement of the vertebræ with tuberculosis include the neural arches and the spinous and transverse processes. A case of isolated tuberculosis of the spinous process of the fourth lumbar vertebra was reported by Anderson, who reviewed the literature in 1940 and encountered 12 similar cases. This lesion was more frequent in adults. Involvement of a single spinous process occurred in 7 patients, and two spinous processes were affected in 5 others. The neural arch was diseased in 5 other cases. Abscess formation appeared, but no muscle spasm or deformity of the vertebral bodies was noted. Anderson stated that these lesions were less common and less grave than those in the vertebral bodies proper. Judd (1940) reported a case of tuberculosis of the transverse process of the third lumbar vertebra in which the destruction of bone was accompanied by obliteration of the psoas shadow on the involved side. Operative intervention disclosed a large abscess from which 4 to 5 ounces of pus were evacuated. A sequestrum the size of an olive was found lying free in the space occupied by the transverse process. Herdner (1950) in a discussion of body section *x*-ray examination as applied to study of the atlas and axis mentioned 3 cases of suboccipital Pott's disease.

Tuberculosis of the sacroiliac joints occurs primarily in young adults, usually associated with tuberculosis elsewhere in the body, the most common sites being the spine and hips. Pain, abscess formation and draining sinuses are frequently present. Pollack and Bosworth (1942) reported that sacroiliac tuberculosis was encountered in 3.4 per cent of a thousand admissions, and described their findings in 44 cases. There were 33 males and 11 female patients varying in age between eight and fifty-five years, with the peak between twenty-one and twenty-five years. In 13 cases the right sacroiliac joint was involved, in 24 the left, and in 7 both sacroiliac joints were diseases. Roentgenographic examination disclosed erosion, sclerosis and cavitation in 73 per cent of the cases. Sequestration could also be demonstrated. The alignment of the joint was disturbed completely in 40 cases, while 9 showed only changes in the lower half of the joint. Changes limited to the central or upper portions of the joint were seen in 1 case each. The sacrum usually showed more apparent destruction than the ilium. Isaacson and Whitehouse (1949) observed spontaneous bony ankylosis of the sacroiliac joint in tuberculosis of this joint. The earliest fusion occurred approximately four months after the onset of symptoms, while the longest interval was approximately two years.

Extension of a tuberculous process into the spinal canal sometimes occurs in the early active phase of the disease. Tuberculous granulomatous tissue may surround the entire cord, separated from it by the dura. Extension of tuberculosis through the dura is rare. Intramedullary tuberculomas of the spinal cord are rare. Key (1940) pointed out that even without mechanical compression the close proximity of tuberculous tissue to the cord may cause toxic and vascular alterations with resultant degenerative cord changes. Even in this type of reaction with no obvious compression, mechanical pressure may be a factor in paralysis. This tends to clear when an abscess communicating with the bony focus and the epidural granulation tissue is drained either by spontaneous rupture or by costotransversectomy. Occasionally a sequestrated mass of diseased bone may be pushed into the spinal canal and compress the cord.

Compression of the spinal cord due to direct extension from a tuberculous pulmonary abscess is rare. Two such cases were described by Norcross (1948). One was that of a thirty-year-old man with tuberculosis of the right lung who developed paralysis with no evidence of Pott's disease.

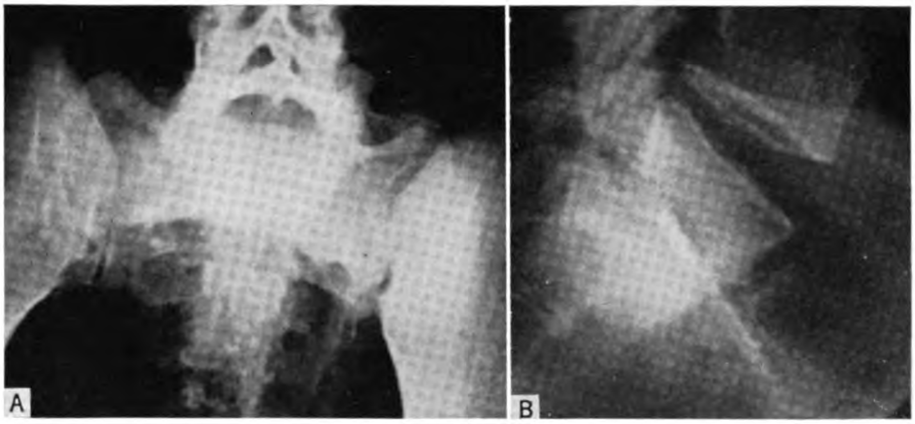

FIG. 133.—*A*, Tuberculosis of the left sacroiliac joint. Note the sclerotic changes in the iliac portion of the joint, and in its midportion a faint rarefied area is visible. *B*, Same case as Figure 133 *A*, lateral view of lumbosacral articulation. Tuberculous destruction of the lumbosacral interspace is present, with a mild forward displacement of the fifth lumbar vertebra. (Courtesy of Dr. N. Pollack, Brooklyn, New York.)

This patient had destruction of the right third and fourth ribs posteriorly with destruction of the pedicles of the third and fourth dorsal vertebræ. At necropsy a cavity was found at the base of the right upper lobe with extension posteriorly through both layers of pleura. From there a sinus tract was found destroying the third and fourth ribs and extending to their pedicles. The infection then entered the spinal canal through the intervertebral foramina causing cord compression. The second case was that of a twelve-year-old boy with pulmonary tuberculosis who developed paraplegia two years after the formation of a cold abscess in the upper thoracic region. Necropsy disclosed tuberculosis of the apex of the left lung. A sinus extended to the back, involving the spinous processes of the third, fourth and fifth thoracic vertebræ, entering the spinal canal through the intervertebral foramina and compressing the cord. Extension of tuberculosis into the cauda equina was described by Alajouanine and Thurel (1945) in a fifty-year-old woman with pain in the lower extremities, buttocks, and lumbar region which had progressed over several years with remissions. Myelography in this patient showed no block, but operation disclosed that the roots of the cauda equina on the left side were enmeshed in fibrous tissue which on biopsy proved to be tuberculous.

A unilateral tuberculous abscess of the thoracic spine with x-ray findings suggesting the presence of a dumbbell neurofibroma was reported by Kneidel, Smith and Bishop (1950). This patient had slight widening of the intercostal space, erosion of the articular tubercle of the right fifth rib and of the transverse process of the fifth dorsal vertebra, with a right paravertebral soft tissue mass due to a tuberculous abscess. The vertebral body was intact. Myelography revealed a complete block at this level, and operation showed caseous material at the level of the fifth and sixth spinous processes. Granulation tissue over the anterior and posterior surfaces of the dura was encountered, and a large tuberculous cavity extended anteriorly and laterally to the fifth and sixth dorsal bodies, conforming with the soft tissue shadow noted radiologically.

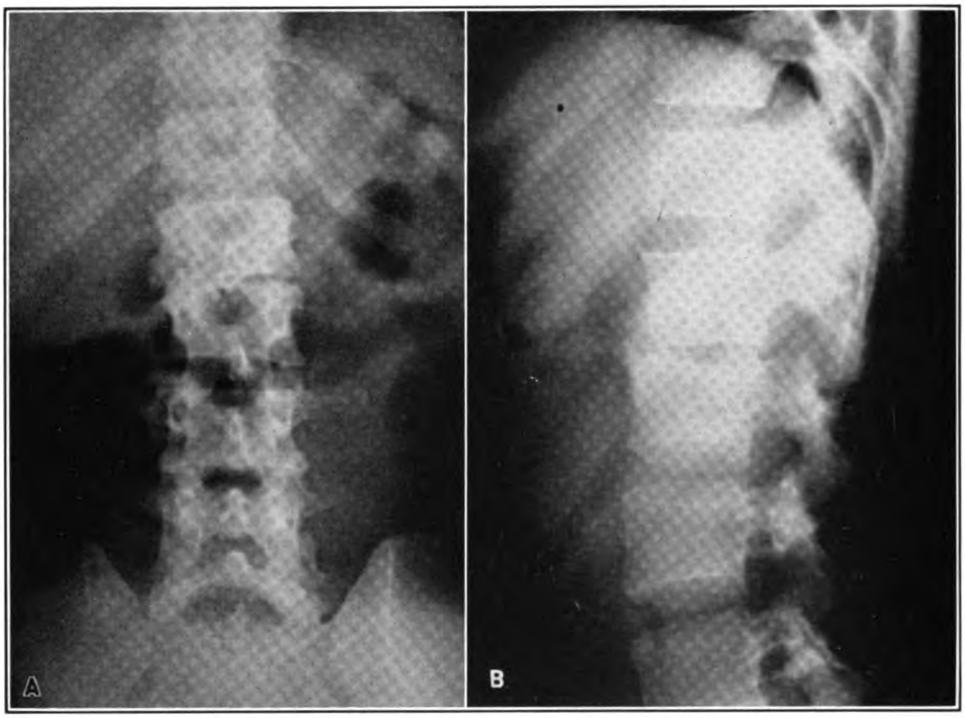

FIG. 134.—Tuberculosis involving the first and second lumbar vertebræ, with irregular narrowing of the intervertebral disc and destruction of adjacent bone, particularly of the superior aspect of the second lumbar vertebra. Slight bony reparative changes are present anteriorly.

Trauma may be a precipitating cause of neurological symptoms in patients with tuberculous scoliosis. In one of the cases reported by Love and Erb (1949) neurological symptoms appeared after a fall in a hockey game. At operation the dura was found to be thickened and adherent to the arachnoid, suggesting trauma with subsequent organization.

A relatively rare form of tuberculosis known as osteitis tuberculosa cystica, occurring as a rule in children, sometimes presents changes in the vertebræ. This condition is usually observed in the diaphyseal aspects of the long bones. It is usually multiple and of metastatic origin from a primary thoracic focus. The disease is insidious in onset, with few or no localizing signs or symptoms. Lesions run a chronic benign course consisting of a circumscribed area of soft tuberculous granulation tissue and degenerated bone which tends to heal slowly. This may occur in a diffuse form in which the cystic area assumes a honeycomb, web-like structure or in a cystic form with destruction of the medulla and cortex. Law (1940) reported a case of the circumscribed type in a six-year-old girl observed over a four-year period with multiple bone involvement. In this case the spine showed circumscribed areas of rarefaction in the third and fifth lumbar vertebræ which healed in about a year.

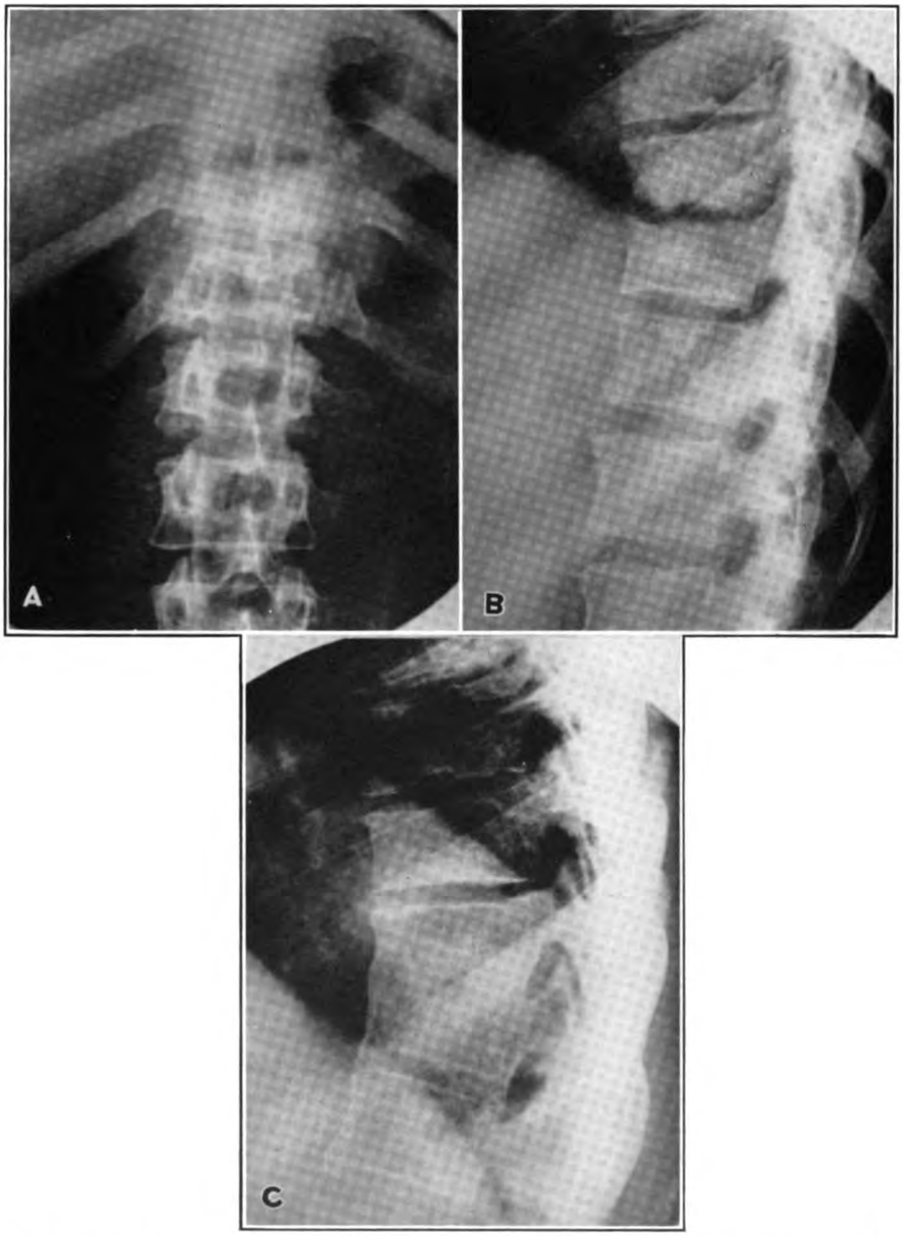

FIG. 135.—*A* and *B*, Tuberculosis of the tenth and eleventh dorsal vertebræ, with considerable osseous destruction adjacent to the intervertebral disc. *C*, Same case, demonstrating healing with bony fusion which occurred under conservative treatment.

The radiologic changes incident to tuberculosis of the spine are very slight in the early stages of the disease. The lesion rarely is diagnosed within six months of the onset of symptoms. The earliest radiologic alteration encountered is a slight decalcification of the superior or inferior aspect of a vertebral body. Less frequently there may be a decalcification of the central portion of the vertebra. Both these changes are extremely difficult to evaluate. Indeed, Hellstadius (1946) reported a case in which an entire vertebral body was the seat of tuberculous necrosis in which the roentgenographic examination was considered to be within normal limits. He mentioned that caseous tuberculosis may spread widely through the medullary spaces without conspicuous injury to the cortex of the bone. Consequently, little apparent change appears on *x*-ray examination. His case also pointed to the fact that the adjacent intervertebral discs are not necessarily involved even if considerable bony change has occurred. As a rule, however, the bony changes are soon accompanied by alterations in the thickness of the adjacent intervertebral fibrocartilage. The sharp line of definition between the bony surfaces and the intervertebral disc becomes hazy. Progressive narrowing of the intervertebral disc follows extension of the infection into this area. With the appearance of areas of erosion partial collapse of the vertebral bodies takes place, becoming progressively more advanced as the disease further destroys bone. Extension of the infection into the adjacent soft tissues is manifested by the development of paraspinal abscesses. In the cervical area the soft tissue extension of tuberculous infection spreads from the anterior aspects of the vertebral bodies, pushing the trachea and esophagus forward. In the dorsal region these manifest themselves as a paraspinal unilateral or bilateral globular or fusiform bulging mass which may displace the paraspinal pleural lines when these are visible. It should be recalled that other diseases such as primary or metastatic

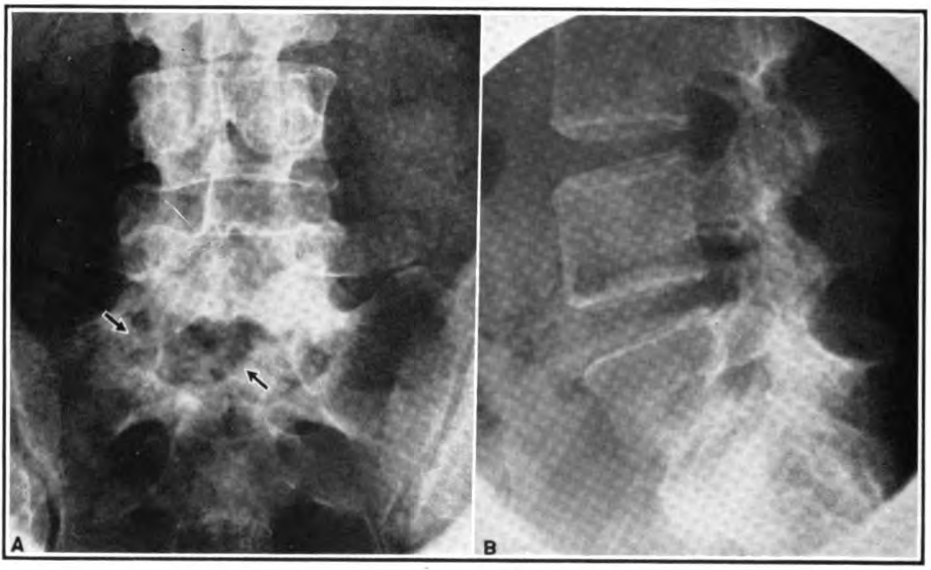

FIG. 136.—*A*, Tuberculosis of the lumbosacral joint. A lytic process is seen in the superior aspect of the first sacral vertebra (arrows). *B*, Same case, lateral view. Note the destruction of the lumbosacral articulation.

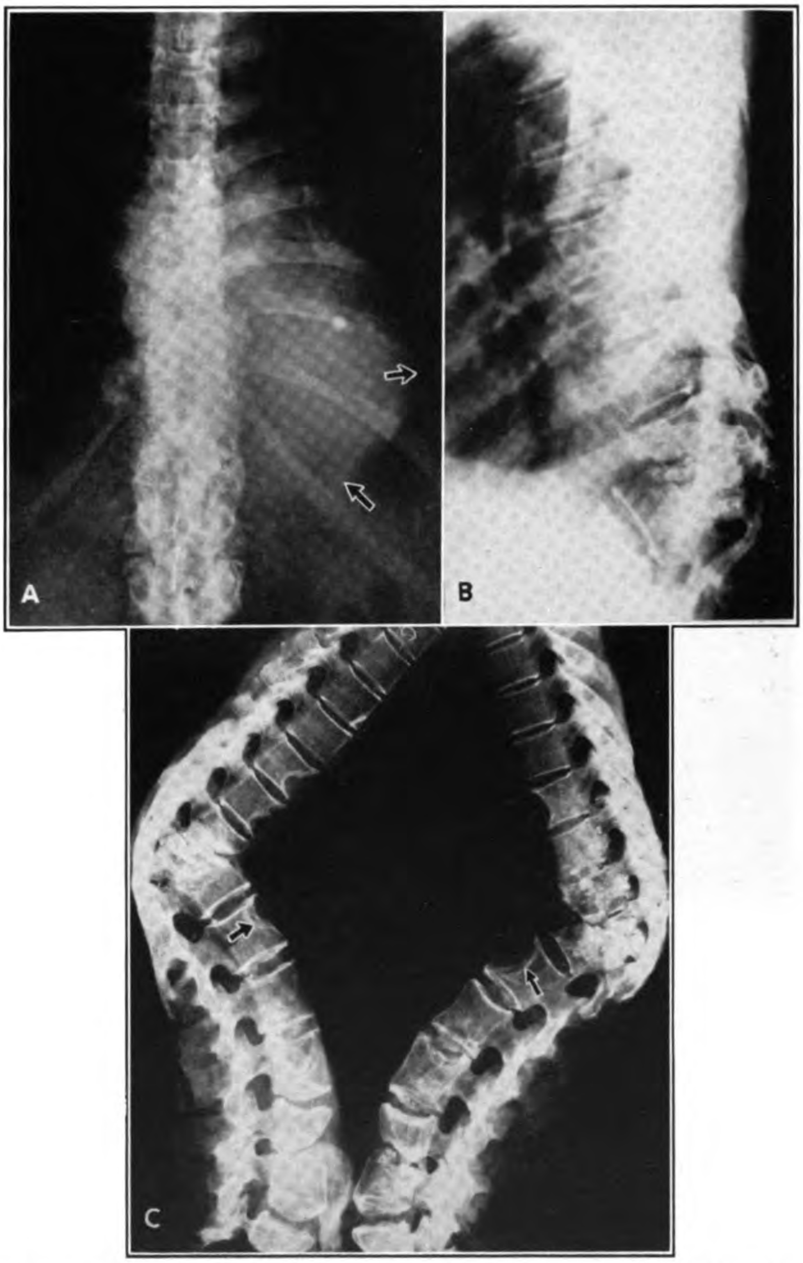

FIG. 137.—*A*, Paraspinal tuberculous abscess of lower thoracic and upper lumbar spine bulging on the left side (arrows). A small calcified paravertebral abscess is seen on the right between the heads of the eleventh and twelfth ribs. *B*, Same case, lateral view. Extensive destruction of lower thoracic vertebræ with gibbus formation. *C*, Same case as *A* and *B*. Specimen of spine sawed in half. Note the destruction in the anterior aspects of the vertebræ immediately above and beneath the gibbus (arrows). The lumbar vertebræ also are affected.

vertebral neoplasms, osteomyelitis, or occasionally paraspinal fluid collections at times simulates this appearance.

Lumbar tuberculous osteomyelitis accompanied by extension of the infection into the psoas muscle produces a bulge on the involved side. In more advanced cases the psoas shadow may be obliterated. In all of these sites it is possible for healing to take place, often manifested by recession of the swelling together with the appearance of calcification within the abscess. These sometimes are very faint amorphous deposits which with progressive healing may become progressively quite dense.

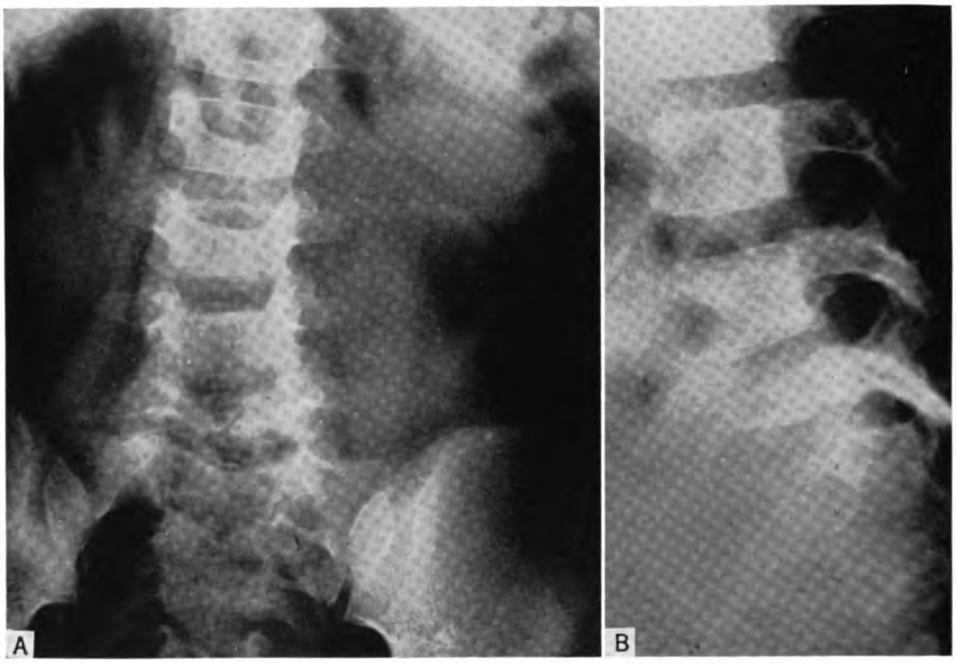

FIG. 138.—Large left psoas abscess in a five-year-old child with tuberculosis of the lumbosacral articulation. Note the bulging psoas shadow on that side. The lateral projection shows flattening of the fifth lumbar vertebra. Destruction of the body of that vertebra is also seen on the anteroposterior view.

In addition to usual roentgen techniques employed in examination of the spine, considerable additional information is obtainable by the use of body section radiography. This is particularly helpful in identifying the extent of destruction of vertebræ in regions of kyphosis, in which the vertebral bodies overlap each other so that they cannot be distinguished one from another. Body section roentgenograms are useful in identifying areas of increased bone density, often revealing small foci of densely ossified bone within areas which are considered to be mainly destructive in nature. These bits of bone represent practically dead osseous tissue. Biopsy taken from the involved area has also been advocated for diagnosis.

Another change sometimes observed with tuberculosis of the vertebræ is a shallow excavation of the anterior aspect of an involved vertebral body. This represents extension of caseation under the anterior spinal ligament producing almost scalloped, sometimes sharply defined semicircular indentations in the anterior aspect of the involved vertebra. This may extend for three or four vertebræ above an area of kyphosis as indicated in Figure 136.

With the onset of healing, increased bone density occurs, and sometimes spontaneous fusion of the vertebral bodies is noted. Not infrequently adjacent paraspinal abscesses develop dense and extensive calcific shadows.

It is often remarked how an extensive deformity of the spinal column may be accompanied by little if any evidence of central nervous system disturbance. However, this does occur, and more often than is generally realized. Key (1940) reported that paraplegia was seen in about 12 per cent of cases, usually in relationship with early active Pott's disease in

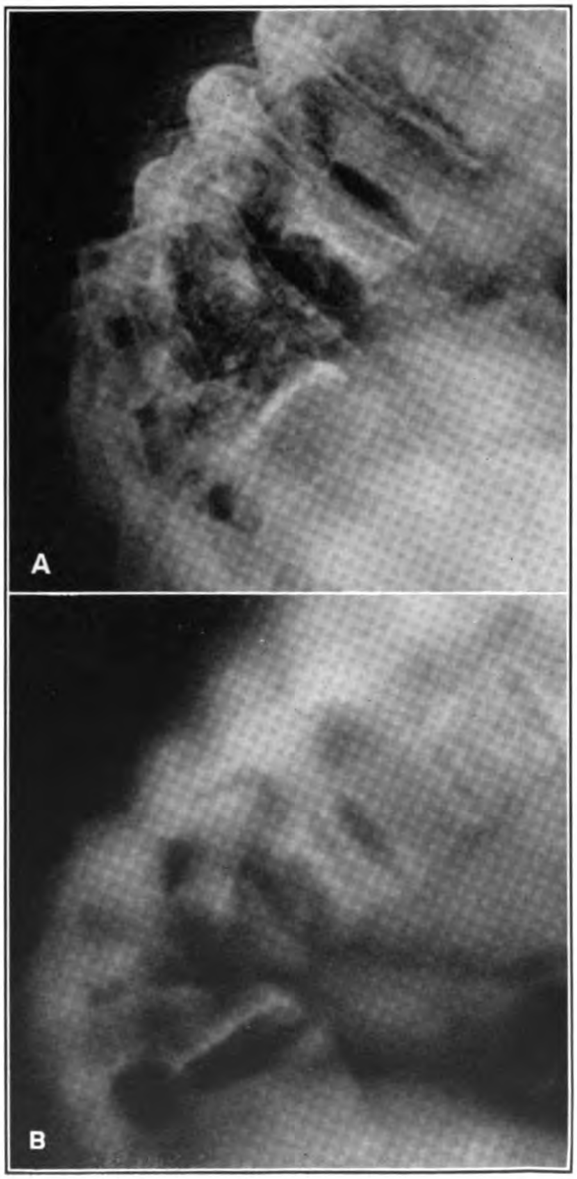

FIG. 139.—*A*, Lateral thoracic spine roentgenogram showing gibbus formation with extensive tuberculous destruction. *B*, Same case, lateral body section roentgenogram.

which there was extension of infected material into the spinal canal producing compression. This may subside as the infection itself recedes. In a review of 30 cases of tuberculosis of the lower spine, Brodin (1951) reported on myelographic observations with water-soluble contrast mediums. Twenty-six of the cases were considered unhealed and 4 were considered healed. In 25 of the unhealed cases myelographic examination showed a varying sized anteroposterior indentation towards the dura in or near the

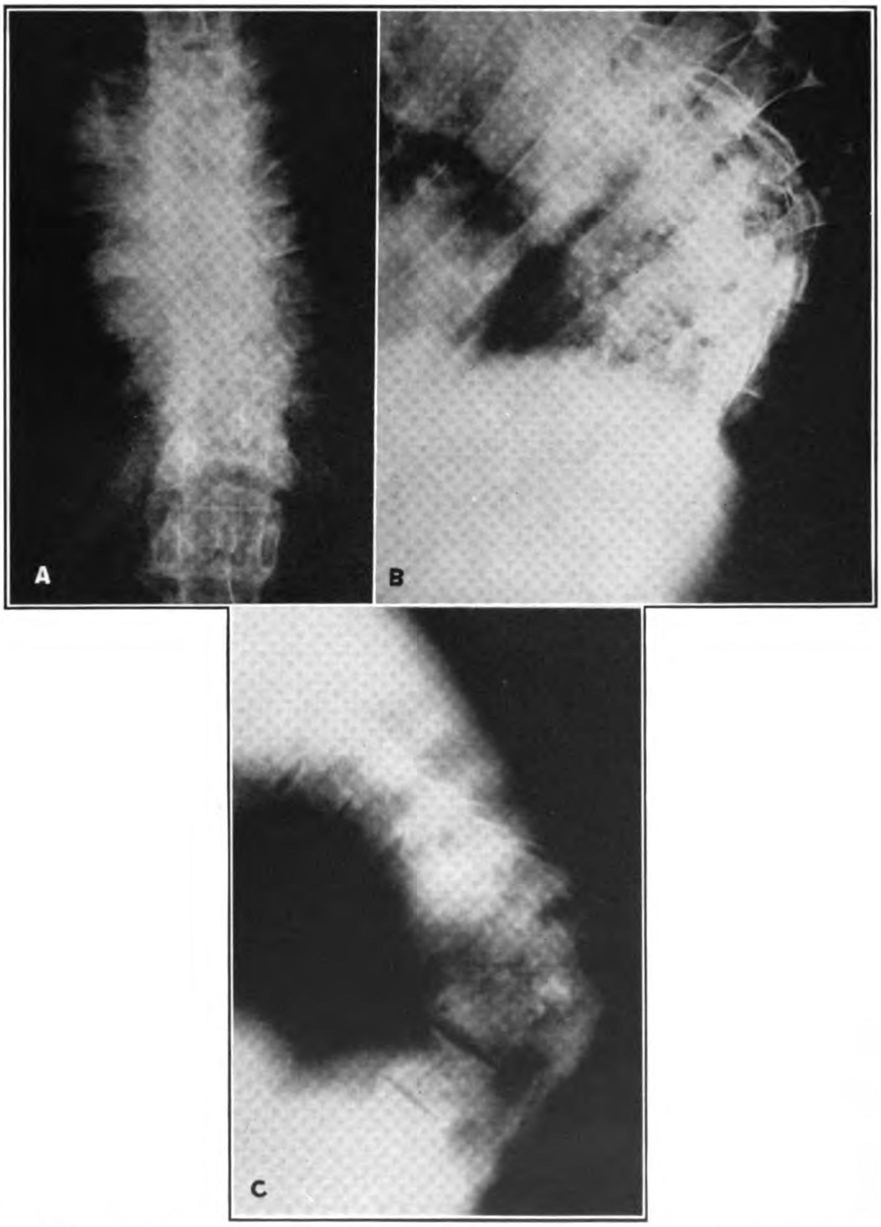

FIG. 140.—*A*, Bilateral tuberculous paraspinal abscess. *B*, Lateral view showing gibbus involving middle and lower thoracic vertebræ. Note the small punctate islands of increased bony density which are situated mainly anterior to the gibbus. *C*, Lateral body section roentgenogram.

midline. There was no correlation between the size of the indentation and that of the skeletal focus.

Our own experiences with myelography in tuberculosis of the spine is relatively limited. One case was of great interest in that the patient was admitted with a clinical history suggesting a protruded intervertebral disc. Preliminary *x*-rays of the spine had been considered normal. Myelographic examination showed a filling defect at the interspace between the

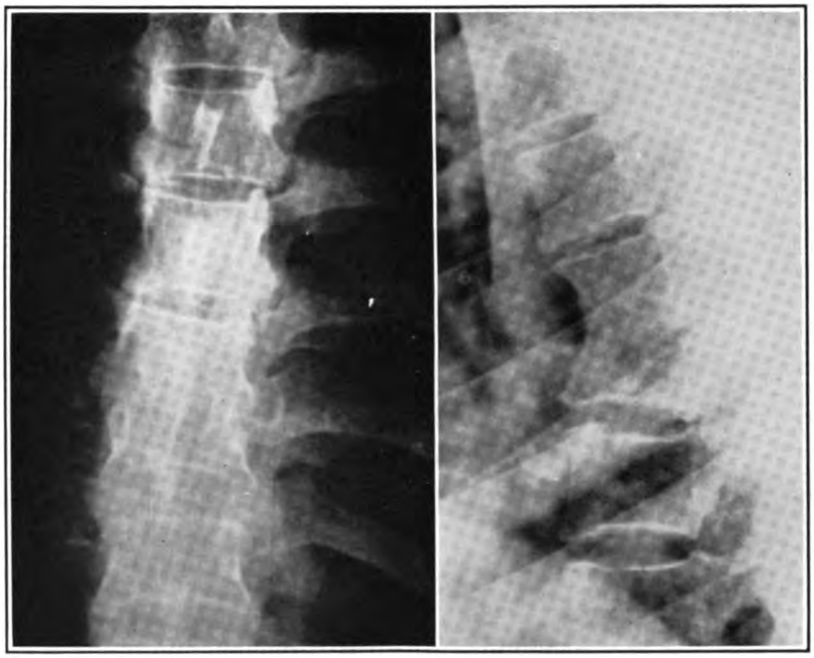

FIG. 141.—Healed tuberculosis of the midthoracic spine. The vertebræ are fused. Note the anterior concavity in the involved vertebral bodies on the lateral film.

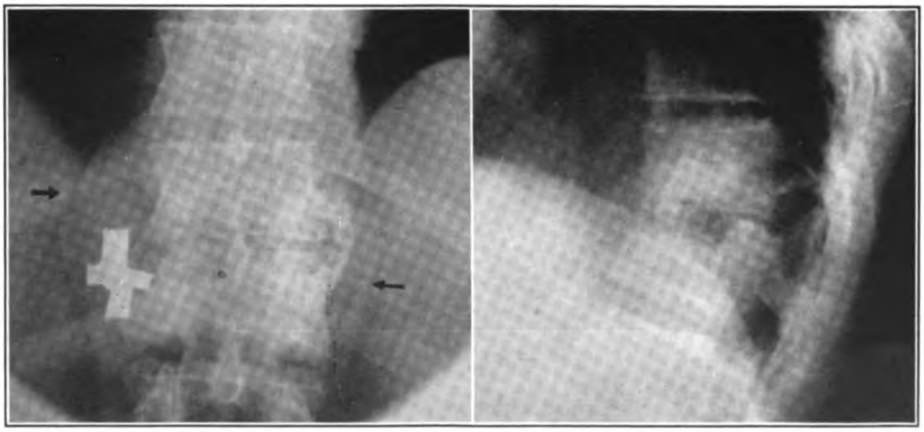

FIG. 142.—Anteroposterior and lateral roentgenograms of eleventh and twelfth thoracic vertebræ with tuberculosis and bilateral paraspinal abscesses (arrows). A large bony spur is present on the left side. Note the destruction of the superior aspect of the body of the twelfth thoracic vertebra on the lateral film.

fourth and fifth lumbar vertebræ on the right side which was considered quite characteristic of a herniated disc. At operation it was found that the myelographic defect had been caused by an indurated pad of fat which was covered with several large epidural veins. Histologic examination of of this pad of fat disclosed a granulomatous lesion. Subsequently the patient developed a collapse of the superior aspect of the body of the fifth lumbar vertebra and after this he developed a fistulous tract leading to the skin above the spinous process of the fifth lumbar vertebra. Material removed at operation disclosed granulomatous tissue consistent with tuber-

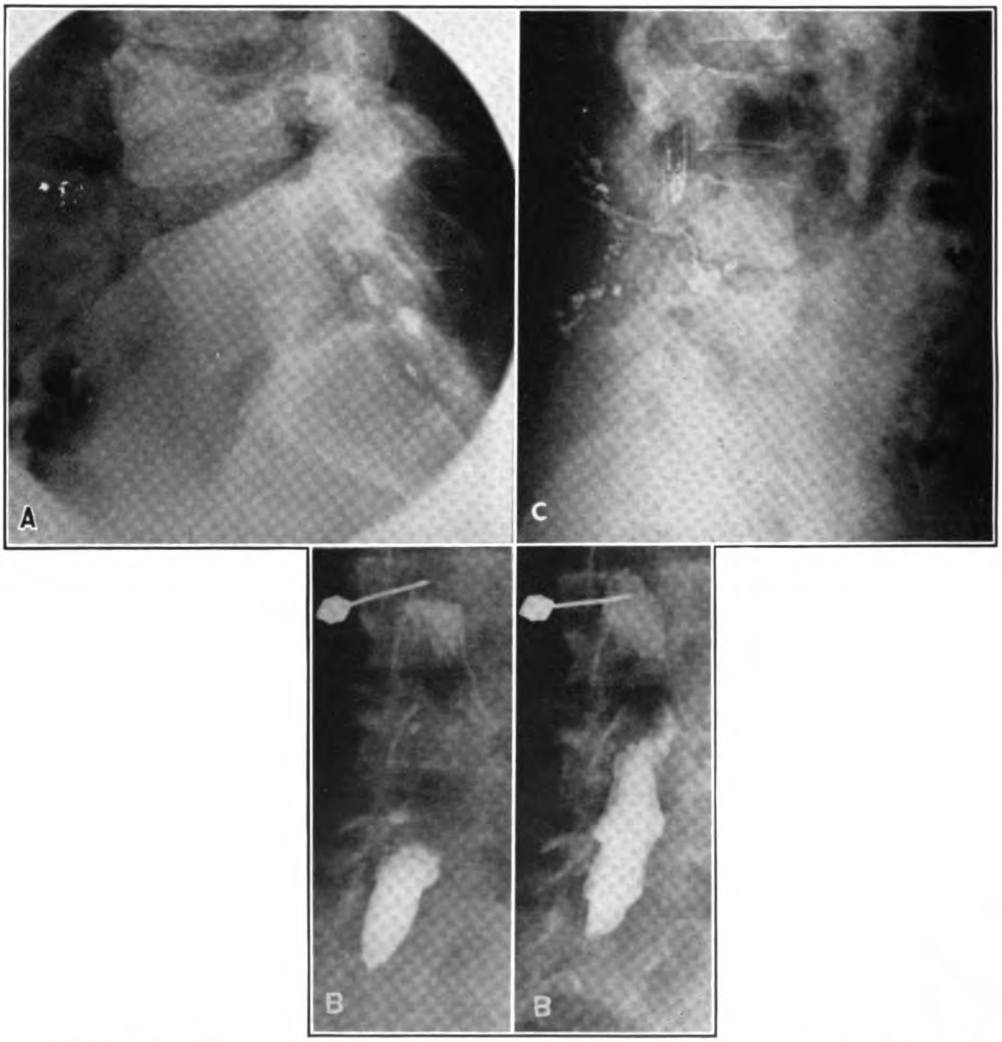

FIG. 143.—*A*, Tuberculosis of the fourth lumbar vertebra in a thirty-five-year-old man with symptoms of cauda equina compression. *B*, The first myelogram in the erect posture shows pantopaque held up at the third lumbar interspace. Note the dentate pattern of the inferior margin of the upper pantopaque. The next myelogram was made in the head-down position. The inferior margin of the intraspinal protrusion is delineated as corresponding with the lower aspect of the destructive process in the fourth lumbar vertebra. *C*, A draining sinus persisted five months after operation. This was injected with lipiodol, and led to the posterosuperior aspect of the body of the fourth lumbar vertebra.

culosis. In another instance of slowly developing paraplegia in a patient with a kyphosis of the mid-dorsal area a complete block at the apex of the kyphosis was observed on myelographic examination.

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Sarcoidosis.—Sarcoidosis is a systemic granulomatous disease of unknown origin. Its pulmonary manifestations are well known, as are the relatively infrequent osseous changes in the small bones of the hands. More often the condition involves the lymph nodes, liver and spleen, the skin and the eyes. The underlying pathologic lesion is an epitheloid cell granuloma or tubercle which heals primarily by fibrosis and scarring without undergoing caseation. The systemic manifestations are referable to the extent of involvement of the various organ systems to a variable degree. Rarely do the osseous forms cause appreciable distress, except for some arthralgic pain.

Involvement of the vertebral column is uncommon. Nevertheless, cases in which lytic changes are present have been recorded. In the patient described by Rodman, Funderbuck and Myerson serial films revealed a rapid lysis which improved after spinal fusion. Wood and Bream summerized five cases, including one of their own, in which the sarcoidosis affected mainly the perivascular spaces in the meninges, the arachnoid becoming infiltrated with lymphocytes and granulomatous nodules attached to the adventitia of the small vessels. Similar perivascular infiltrations occur within the spinal cord, the destruction of nervous parenchyma being consequent thereto. The myelitis and meningitis may be indistinguishable from an intramedullary tumor both radiologically and at operation, and arachnoiditis likewise may be simulated.

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SPONDYLOSIS (HYPERTROPHIC OSTEOARTHRITIS, OSTEOPHYTOSIS, BECHTEREW'S DISEASE, SPONDYLOSIS DEFORMANS)

The many names applied to this common condition offers a subject for conversation. It is commonly recognized as osteoarthritis, but the term is a misnomer because no definite inflammatory element is associated with the disease. Another name applied to the condition is Bechterew's disease, but Oppenheimer pointed out that the original reports were so ambiguous that Bechterew's disease has come to imply in English and French a certain type of senile kyphosis, while in the German and the Scandinavian literature Bechterew's disease is considered another name for what we designate as ankylosing spondylitis. It is Oppenheimer's (1942) opinion, with which many agree, that the term Bechterew's disease be dropped, since it defines neither a clinical nor an anatomic entity. Borak (1947) preferred the term marginal spondylosis if spurs alone were present at the edges of the vertebral bodies. The term destructive spondylosis was proposed if spurs coexisted with narrowing of an intervertebral space. He further suggested that spondylosis may be a factor in the development of spondyloarthritis because of the frequency with which they occurred together. Collins points out that the suffix "sis" or "sos" should not have been applied to a normal anatomical term like "spondylo," which means simply vertebra. He suggested that the term "osteophytosis" would be more appropriate, but in view of the wide acceptance of the designation "spondylosis" the concept be that this refers to the clinical syndrome arising from the effects of discal degeneration and consequent hypertrophic spurring and deformities of the intervertebral spaces and foramina.

Arthritis of the vertebral column occurs in the true joints lined with synovial membrane, including the apophyseal and costovertebral articulations. Inflammatory reactions in these can produce painful symptoms. Malalignment of the vertebral bodies incident to degenerative changes in the intervertebral discs also can produce changes in the alignment of the various synovially lined joints, with consequent irritation, reactive response to trauma and inflammation. In the early stages little visible change occurs, but later there is thickening of the facets, narrowing of the joint spaces, irregularity of the articular surfaces and finally obliteration of the joint and ultimately ankylosis. Concomitantly varying degrees of spondylosis exists, and the combination of the two processes is frequent. The compressive effects of spondylotic ridging often is heightened by the thickening of the laminæ and ligamenta flava associated with arthritis. In spinal canals with narrowing at their lateral recesses, particularly those with narrowed sagittal diameters in the lowermost two lumbar segments, neurologic and nerve root symptoms out of proportion to the spondylosis or small discal protrusions are often encountered.

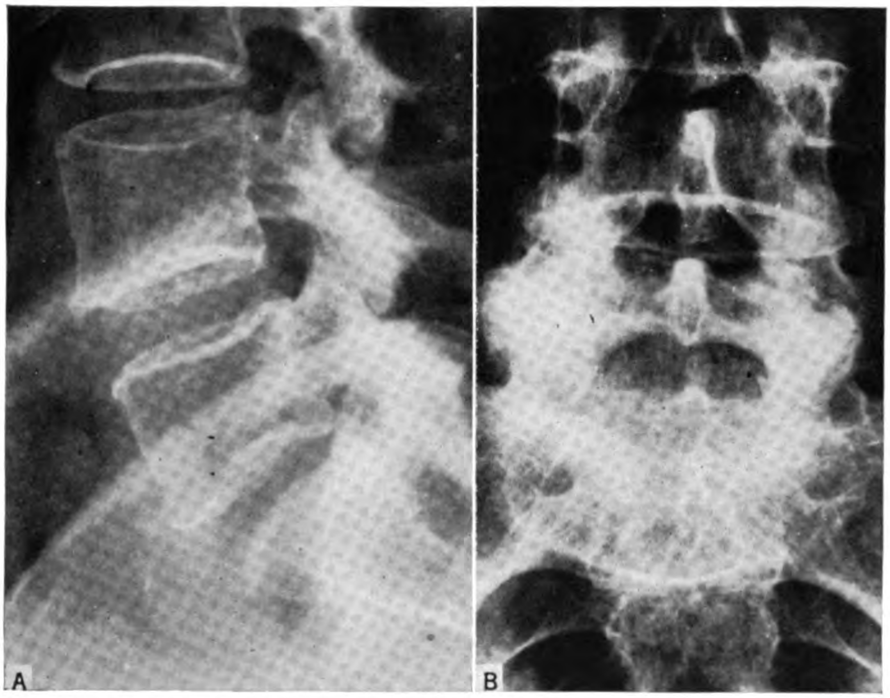

FIG. 144.—Arthritic changes involving the apophyseal joints between the fourth and fifth lumbar vertebræ and the fifth lumbar and first sacral vertebræ. Note the heavy sclerotic changes.

In general spondylosis is a degenerative condition which usually begins in early middle age. Most of the time it is asymptomatic unless the spondylotic deformities produce pressure on the spinal cord or nerve roots. The primary change of the disease occurs in the intervertebral discs (Horowitz, 1940). With the progressive loss of turgor and elasticity incident to advancing age there is variable softening and weakening of the disc margins. The softened cartilages swell outward, bulging the annulus fibrosus. These then serve as bridges over which periosteum may extend with consequent formation of osteophytes. In the early stages of the disease the degree of calcification of the bridging osteophytes is minimal, and hence largely radiotransparent. As more and more calcium is laid down and bone is formed, the spurs become prominent. Involvement of the cervical spine is quite frequent, and may precede similar spondylotic changes in the thoracic and lumbar regions. The sacroiliac areas as a rule are not involved, in contradistinction to the frequency of such change with rheumatoid spondylitis. The spondylotic ridges may extend around the periphery of the intervertebral joints, and protrude into the spinal canal and the intervertebral foramina. Narrowing of one or more intervertebral discs may develop together with changes in the curvature of the spine. The dorsal spine may assume a rounded appearance. The usual lumbosacral lordotic curve may become straightened and the spine tilt to one side or the other.

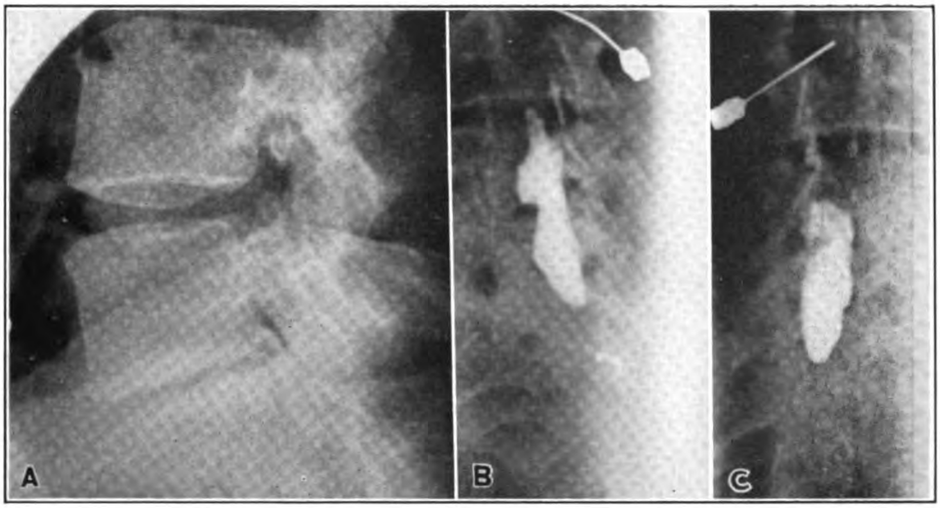

FIG. 145.—*A*, Lateral conedown roentgenogram of lumbosacral articulation. A spur is seen protruding dorsally (arrow) from the posteroinferior aspect of the fifth lumbar vertebra. The lumbosacral interspace is narrowed. *B* and *C*, Right and left oblique myelograms showing indentation of the pantopaque column. The defect was found at operation to be due to a heavy spur corresponding to that seen in Figure 145 *A*.

As a rule the development of posterior discal spurs is less extensive than in the periphery. Keyes and Compere (1932) showed experimentally that fibrillation of the cartilage plate as a result of chronic trauma led to eventual dehydration and escape of disc material of microscopic size. Nevertheless, this resulted ultimately in loss of the disc's semi-fluid consistency and of its cushioning effect. As the disc became thinner, the daily trauma of living was borne in greater part by the fibrocartilaginous annulus fibrosus. A strain was placed upon the vertebral bodies with consequent sclerosis, lipping and spur formation. The preponderance of anterior and lateral osteophyte formation was explained by Keyes and Compere by the shift in the axis of motion of the vertebræ posteriorly to the articular facets when the disc lost its normal configuration. An added stress forward and laterally was produced when the patient bent. This lipping and spurring also occurred in the true arthrodial joints of the articular facets of the spine with consequent bony overgrowth.

The clinical picture associated with spondylosis deformans is usually referable to the area in which the change exists, provided that the pathologic alterations have produced pressure upon the spinal cord or upon the nerve roots. If an inflammatory reaction accompanies the spur formation symptoms may be produced without specific nerve or spinal cord pressure. It is difficult to generalize on the correlation with pain, and one has to conclude that while considerable deformation is known to be without pain in some, others may have considerable pain with similar or lesser deformities. Therefore one must exercise considerable and experienced judgement in each

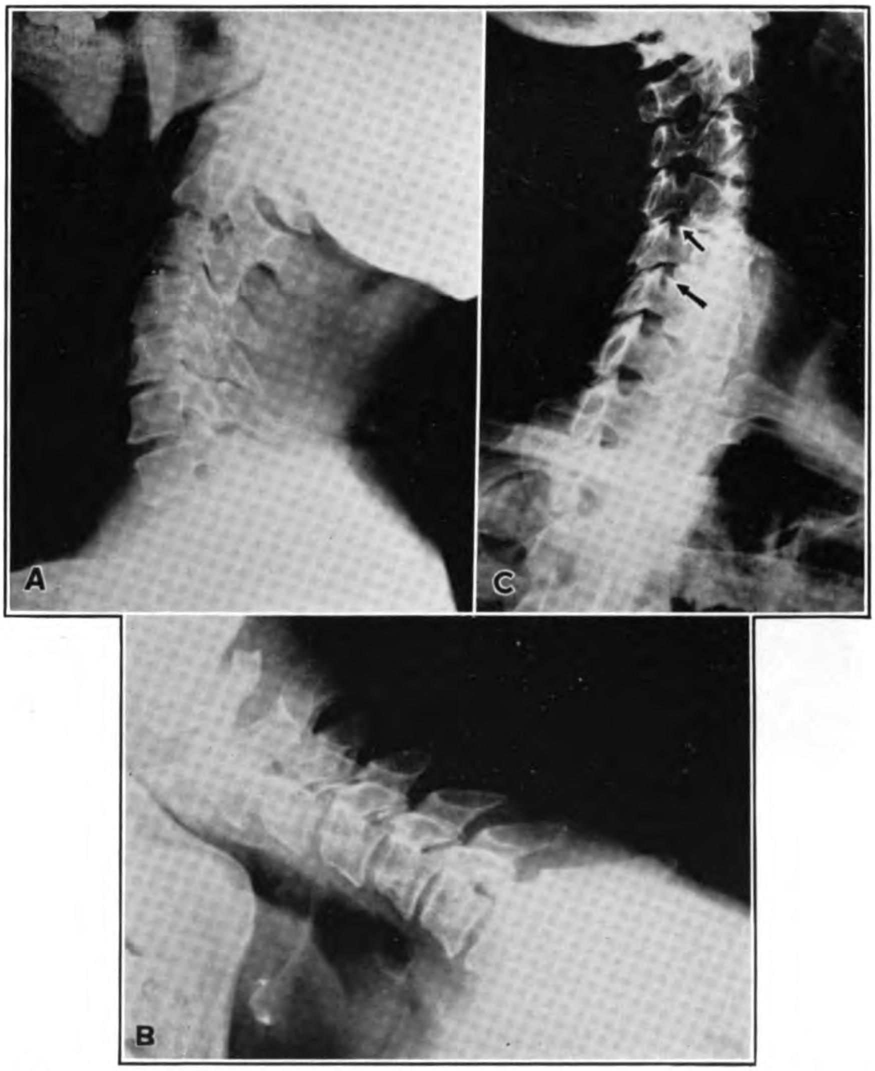

FIG. 146.—*A* and *B*, Extension and flexion lateral spine roentgenograms in case of mild spondylosis with slight narrowing of lower cervical intervertebral discs. *C*, Spurs are noted projecting into the intervertebral foramina between the fourth, fifth and sixth cervical vertebræ (arrows).

situation, judging from the individual patient rather than from a broad conclusion.

Radiologic investigations for spondylosis and arthritis are based on plain films examinations and myelography. The architectural alterations are quite adequately seen on preliminary roentgenograms, but pressure effects on the spinal canal cannot be estimated with myelography. This should not be undertaken unless the clinical picture warrants thought of surgical relief. In examination of the cervical spine anteroposterior, lateral and oblique films are required. Special views of the atlas and axis, and films made in flexion and extension should be made at the discretion of and under the supervision of the responsible radiologist. It is quite possible to identify spondylotic changes on anteroposterior and lateral films, but if impingement into the intervertebral foramina is sought the oblique views are very helpful, particularly when made for stereoscopic viewing. Plain films made in the anteroposterior and lateral projections may suffice in so far as the thoracic spine is concerned. For the evaluation of the condition in the lumbosacral area oblique views for identification of the articular facets may provide additional information. Myelography constitutes a very important element in adequate examination if there is evidence of nerve root or spinal cord pressure. Indeed, it is well recognized that the pressure effect of spurs and ridges projecting into the spinal canal may simulate various degenerative diseases, and it has become common practice to investigate the so-called degenerative diseases myelographically before excluding pressure as a cause for bizarre symptomatology.

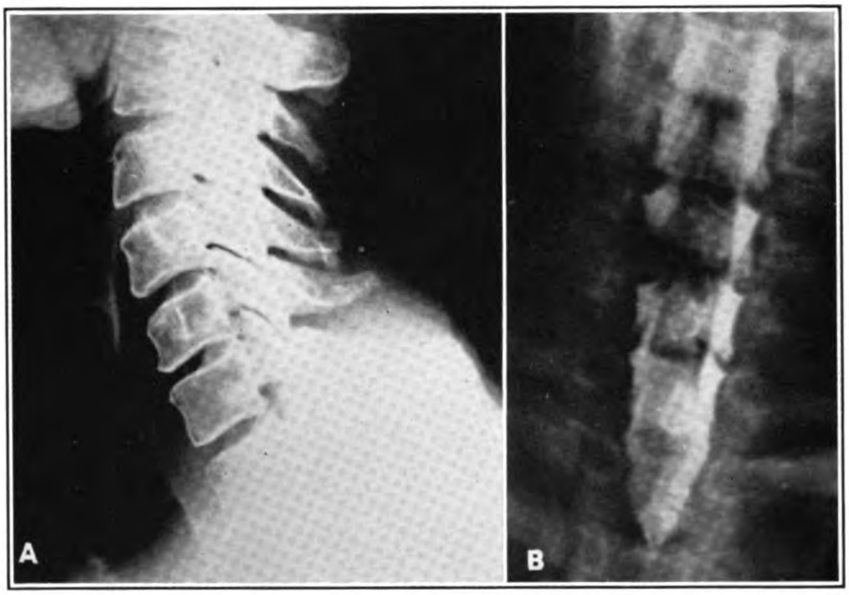

FIG. 147.—*A*, Lateral cervical spine roentgenogram. The vertebral bodies are well formed and aligned. *B*, Cervical myelogram. A semicircular defect is present on the right side at the sixth cervical interspace. At operation this was found to be due to a dense cartilaginous spur. The clinical picture was that of a lateral herniated disc.

Spondylotic deformities may impinge on the foramina transversaria and cause compression of the vertebral artery, thereby producing a clinical syndrome of vascular insufficiency with headache, vertigo, visual and speech defects and gait impairment. This is best demonstrated by means of vertebral artery angiography, which serves to identify intraluminal obstructions as well. The effects of changes in position of the head and neck by rotation or hyperextension in altering the circulation through the vertebral arteries can be produced in some patients by manipulation, and this may be enhanced by the compressive effects of osteophytic changes. In some in-

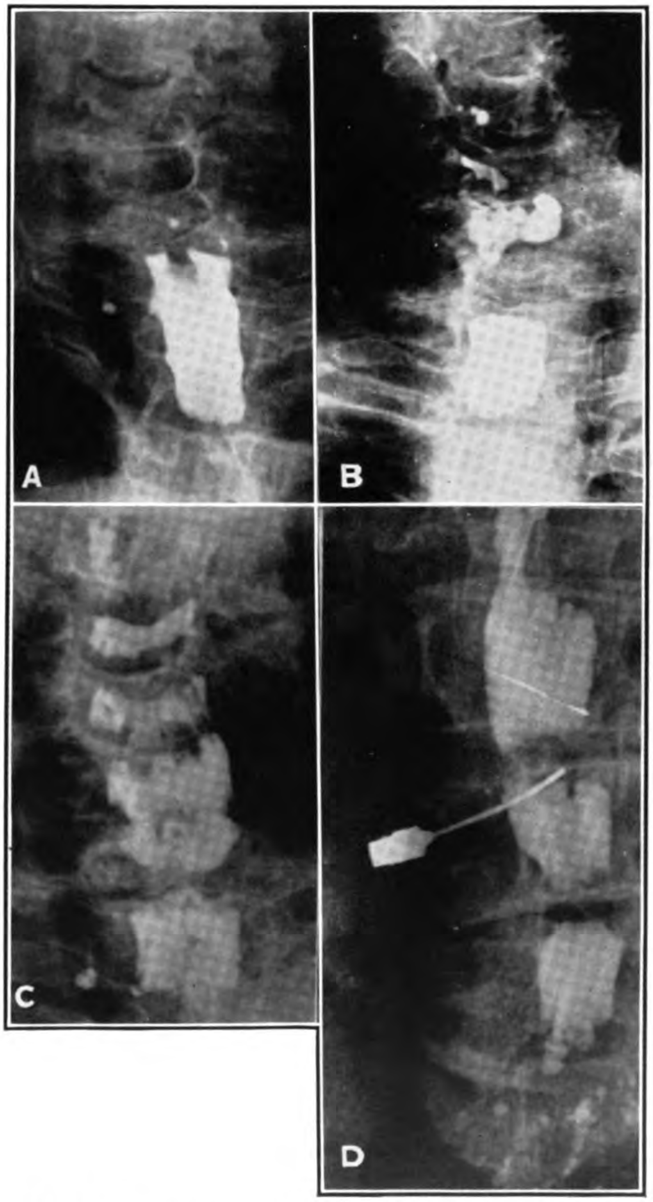

FIG. 148.—*A*, *B* and *C*, Successive spot films made during cervical myelography. At the time *A* was exposed there was a temporary delay at the seventh cervical interspace. A few seconds later a small amount passed above this region (*B*), and within a minute a "wash-board" ribbed effect was present (*C*). This patient had symptoms of cervical spinal cord compression. *D*, Same case. Even though the patient had no symptoms referable to the lumbar spine this region was examined, and radiologically presented the same type of "washboard" deformity.

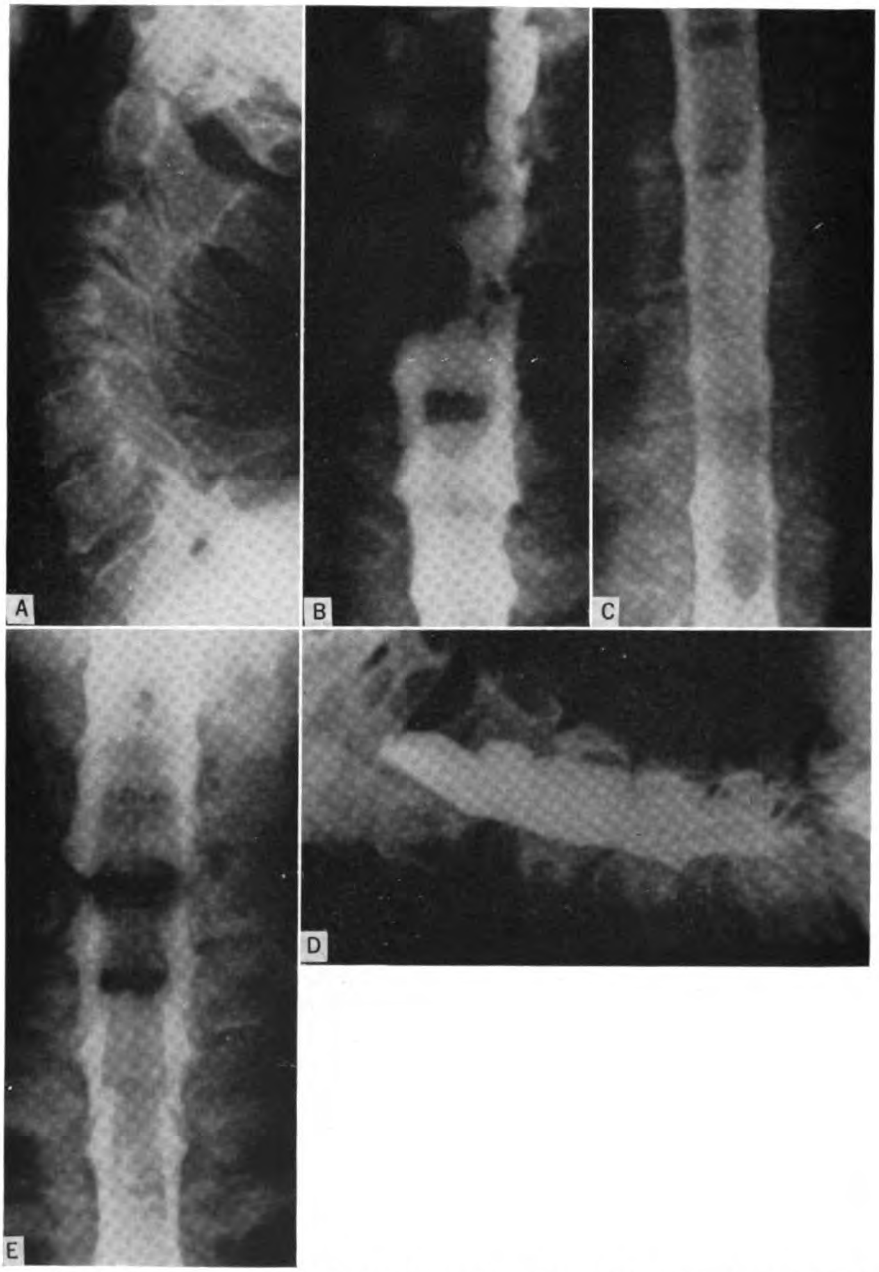

FIG. 149.—A 58-year-old woman with weakness of both arms and progressive spastic gait and weakness of lower extremities for about 4 years. *A*, Lateral roentgenogram of cervical spine shows only mild hypertrophic spondylotic changes. *B*, Myelography showed a partial block at the level of the fifth cervical interspace, with the pantopaque flowing to the right of the spinal canal. *C*, Film of thoracic canal showing the pantopaque backing up in this region. *D*, Transcervial view with head in moderate extension. A large indentation is present in the anterior aspect of the column. Note the upward displacement of that nerve root, as well as the flattening of the cord. At operation a large ridge was present. The laminæ of C-6 were unusually dense and thickened, contributing to the compression of the cord. *E*, After the cervical canal was carefully filled, avoiding passage of pantopaque into the skull, a ridge defect becomes evident at the fifth interspace. The spinal cord is widened. Note the anterior spinal artery in the middle of the canal, with vessels passing laterally at the level of the upper aspect of C-6.

stances dilatation of the vertebral artery is sufficient to produce erosive changes along the foraminal walls, but this is infrequent.

In the cervical spinal canal, myelograms often provide evidence of a localized lesion which may be amenable to surgical relief. From the purely radiologic viewpoint the defects present themselves either in the lateral aspect of the spinal canal, and are demonstrable as filling defects of one of the axillary pouches, most commonly in the last three or four cervical intervertebral spaces. Also of great interest are ridges in the dorsal aspect of the intervertebral discs extending into the anterior spinal canal. Myelographically these manifest themselves occasionally in advanced cases by a complete block at the level of one of these ridges, or a partial block permitting passage of pantopaque beyond successive ridges. Sometimes the myelogram

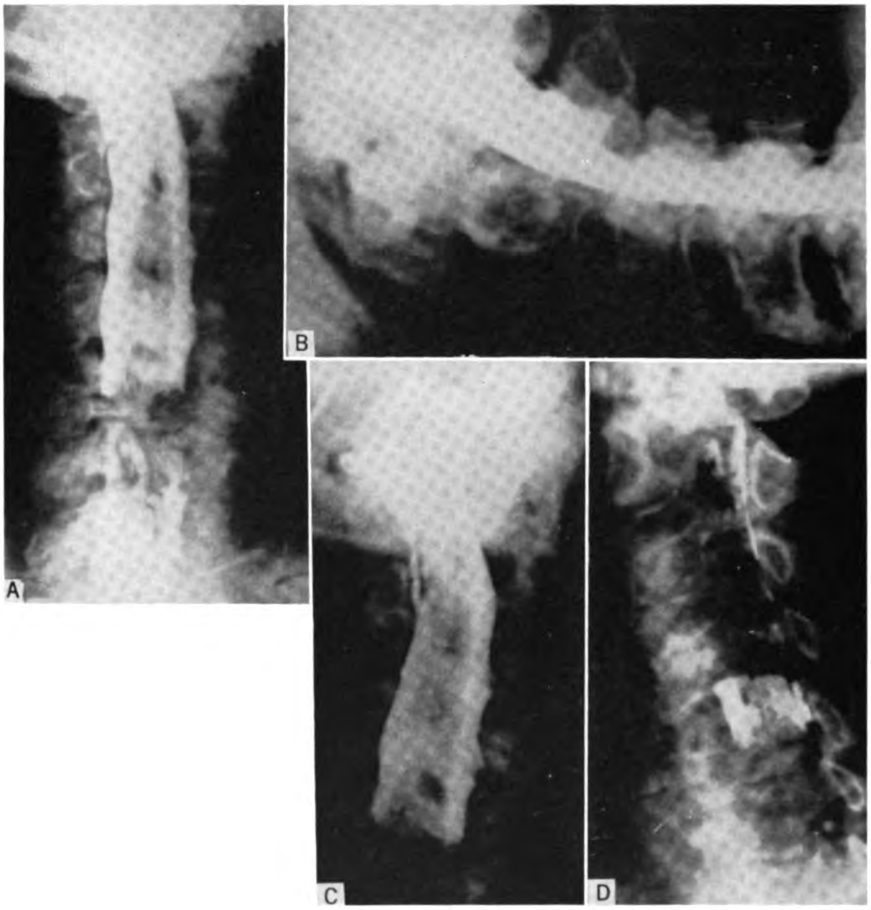

FIG. 150.—A 48-year-old woman with intense pain in the right arm, weakness and paresthesias, and weakness of both lower extremities. *A*, The flow of pantopaque in the cervical canal was held up at the interspace between the fifth and sixth segments. A transverse defect, wider on the right side, is present. *B*, Transcervical view with canal filled. Note the dorsal indentations at the fourth, fifth and sixth interspaces. The column is indented anteriorly at the sixth interspace. *C*, With the patient placed erect, a marked delay is noted in the downward passage of the pantopaque at the sixth interspace. The oil below the column has passed rapidly down the spinal canal. *D*, Film made about three minutes after *C*. Some pantopaque is still held up above the ridge. Verified at operation.

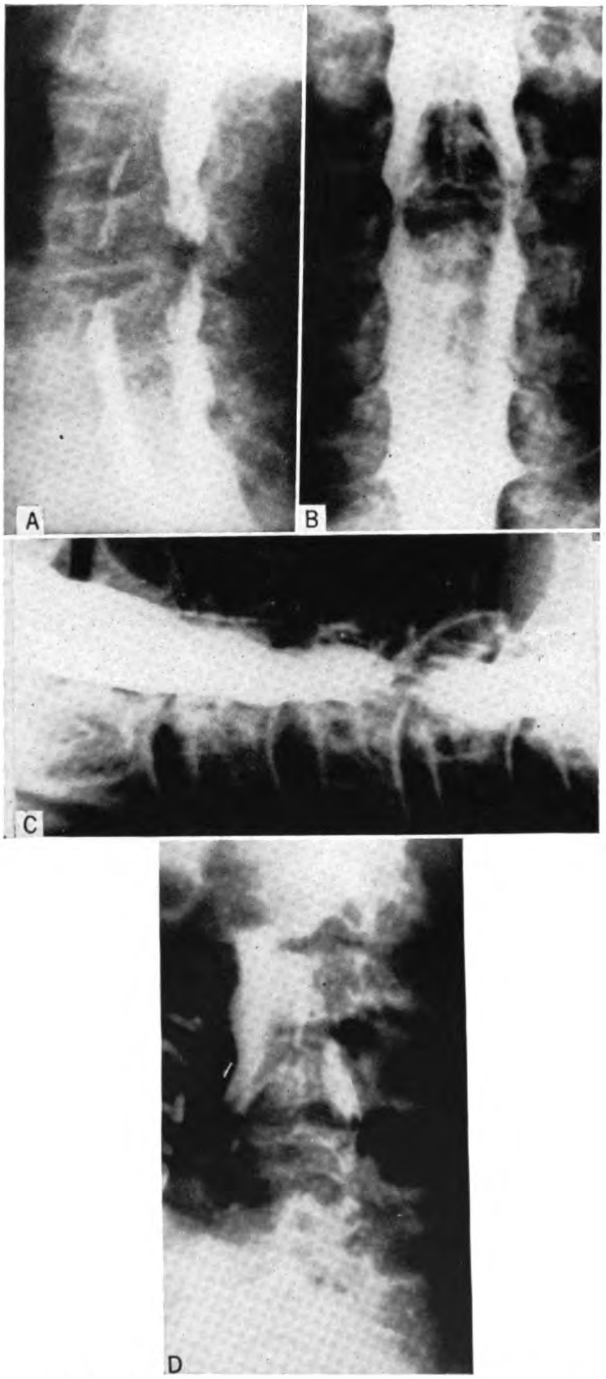

FIG. 151.—A 60-year-old man with a clinical diagnosis of syringomyelia. Numbness and burning both hands and feet, weakness of both arms and legs, broad based gait. Temperature, touch and vibratory sense intact. *A*, A delay in the passage of pantopaque at the fourth interspace was noted. *B*, The column then became spread, producing a spindle-shaped defect which at first glance was believed to be suggestive of an intramedullary tumor. *C*, Transcervical study shows compression of cord from below as well as from above, leading to the diagnosis of a large ridge defect associated with thickening of the neural arch and ligamentum flavum. *D*, When the patient was placed erect the pantopaque below the defect passed rapidly down the spinal canal, but that above was held up, and presented a widened configuration simulating that of a tumor. Verified at operation.

presents an appearance quite suggestive of the rippling of a washboard. In the demonstration of these defects anteroposterior, oblique and lateral views should be available. In obtaining the lateral views some rotate the patient to the side, but a more adequate lateral examination can be obtained by trans-cervical films in which the pantopaque is demonstrated along the floor of the spinal canal. Caution should be exercised in the interpretation of minor defects as seen in the trans-cervical view. On performing this examination routinely in a number of patients who had lumbar herniated discs with asymptomatic cervical spondylosis it was possible to demonstrate indentations into the pantopaque column which certainly could not be held accountable for anything of clinical importance even though the deformities were quite prominent.

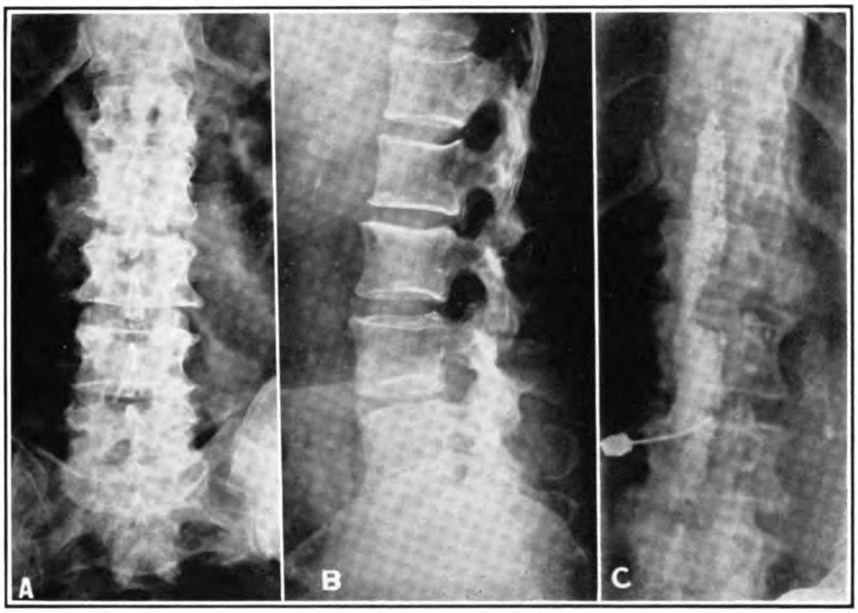

FIG. 152.—*A* and *B*, Anteroposterior and lateral roentgenograms of lumbosacral spine in patient with syndrome of herniated lower lumbar disc. Moderate spondylotic changes and straightening of the usual lumbar lordotic curvature. *C*, Myelograms reveal a semicircular indentation on the left side at the second lumbar interspace. At operation this was found to be due to a large spondylotic spur.

In evaluating the compressive effects of osteophytes on the available space within the cervical spinal canal the sagittal diameter must be taken into consideration. If this is less than 14 mm. relatively small ridges can cause disproportionate pressure effects on the anterior aspect of the cord, especially if there also exists thickening of the neural arches and of the ligamenta flava. It is important to ascertain the degree of deformity produced on the pantopaque column by carefully studying the effects of cautious extension of the head on trans-cervical myelograms. While indentations produced by wrinkling of the ligamenta flava may have no specific significance in patients with wide spinal canals, they can be of considerable importance when the sagittal diameter is diminished.

The coexistence of spur formation in the lumbar spinal canal with herniated discs is by no means uncommon. In our own experience we have encountered approximately 40 cases in the past five years in which spurs produced filling defects which were considered indistinguishable from herniated discs when they occurred in the lower two lumbar interspaces, and spinal cord tumors when they occurred higher. The coexistence of spurs in one interspace with protruded intervertebral disc in another interspace was encountered fairly often. Spurs alone also simulate herniated discs to such an extent that one cannot be distinguished from the other until operation. Indeed, the relationship between uncovertebral osteophytes and discogenic disease is so close that the latter is regarded a forerunner of the development of spur formations.

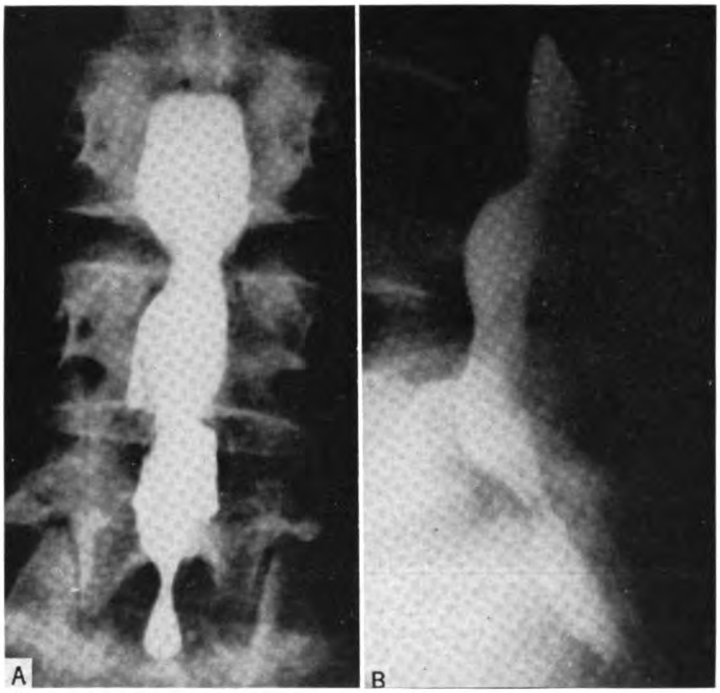

FIG. 153.—Large volume erect myelogram, 44-year-old woman with pain in both lower extremities as well as low back pain. Large spondylotic spurs indent the anterior aspect of the pantopaque column, and deform the lateral aspects as well.

The effects of ridging and osteophytes in causing compression on the cauda equina is frequently accentuated by concomitant arthritic changes in the apophyseal joints. Heavy bony thickening of the laminæ and hypertrophy of the ligamenta flava can sufficiently diminish the available space in the spinal canal to greatly exaggerate the pressure effects of relatively small discal protrusions, ridges, osteophytes or spondylolisthesis. In this regard the anatomic structure of the pedicles of the lowermost three lumbar vertebræ is important. At the base of the pedicle in its cephalad and caudad aspects one can identify a smooth, groove-like configuration forming the inferior and superior aspects of the intervertebral foramen. Dr. Joseph A. Epstein has observed at operation that the superior aspect of the involved pedicle not infrequently is transformed into a narrow, groove-like structure by bony overgrowth, and that this may have a marked compressive effect on the contained nerve root. In some, it is believed that a developmental defect predisposes towards this narrowing because the sagittal diameter of the canal also is diminished. A resemblance to the narrow spinal canal seen in achondroplastic dwarfs has been noted in some cases. In others, where the canal is of normal configuration and dimension, it is considered that the deformity is part of an acquired, degenerative process. In either event, the chance of success of surgery is enhanced by adequate unroofing of this constricting malformation. Spur formations also appear on the superior articular facet of the subjacent vertebra, but these are less likely to be troublesome.

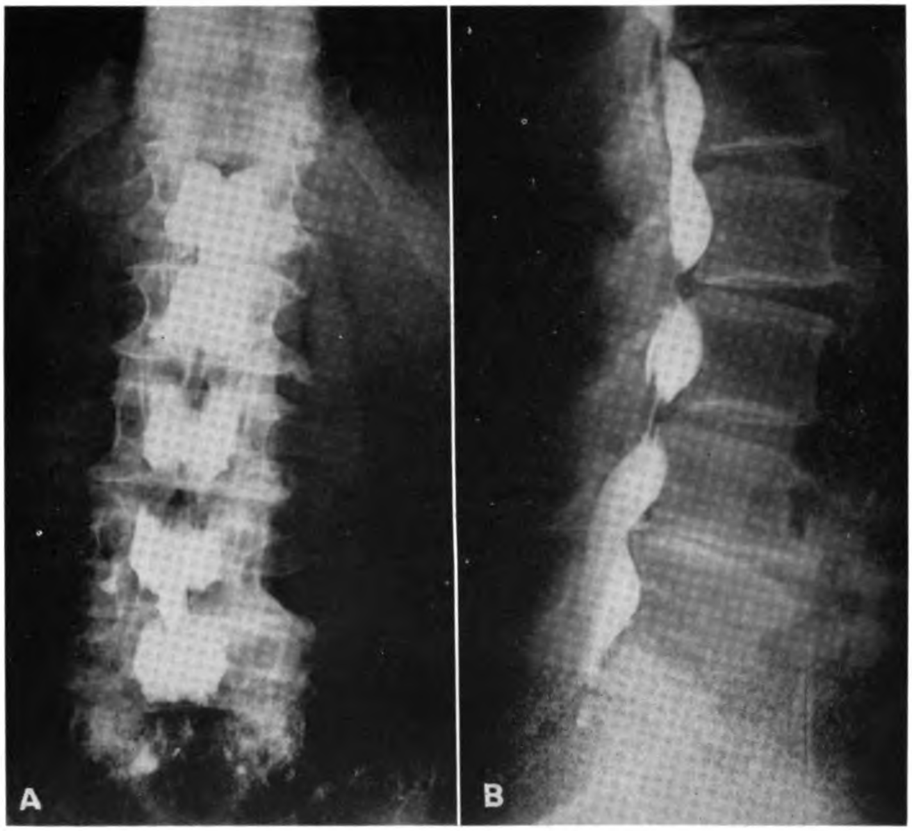

FIG. 154.—Anteroposterior and lateral myelograms of a 59-year-old man completely incapacitated because of pain and weakness of both lower extremities. Large ridge defects are present at each lumbar level. Note the narrow configuration of the pantopaque column at each discal space produced by the bulging at the disc as well as pronounced thickening of the neural arches and ligamenta flava. Considerable relief after multiple laminectomies. (Courtesy of Dr. Joseph A. Epstein.)

In a review of our material we have noted that it is very difficult to identify these changes radiologically. Sometimes the recognition of a narrowed sagittal diameter of the lower lumbar spinal canal has been helpful. We accept a depth of 1.5 cm. as the approximate lower limit of normal, measuring from the floor of the spinal canal to the inferior margin of the superior articular facet. The effects of hyperextension on the available space in the lumbar spinal canal can best be demonstrated myelographically. It is necessary in these studies to fill the lumbar canal up to about the third

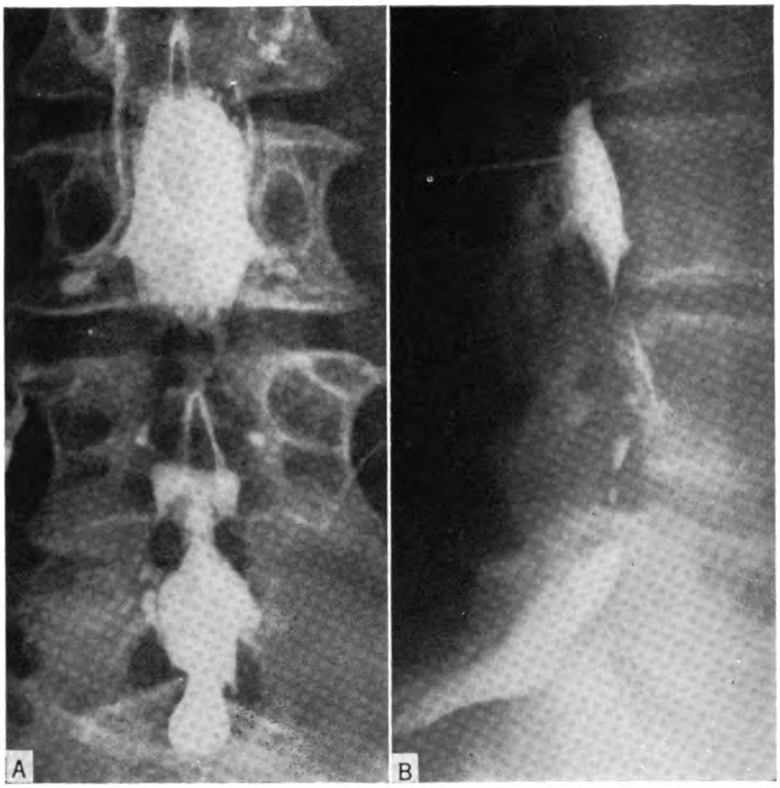

FIG. 155.—Pronounced narrowing of the sagittal diameter of the spinal canal produced by discal ridges anteriorly with markedly thickened neural arches and heavy ligamenta flava. At operation the neural arches were almost stony hard. (Courtesy of Dr. Milton Elkin, Albert Einstein College of Medicine, New York.)

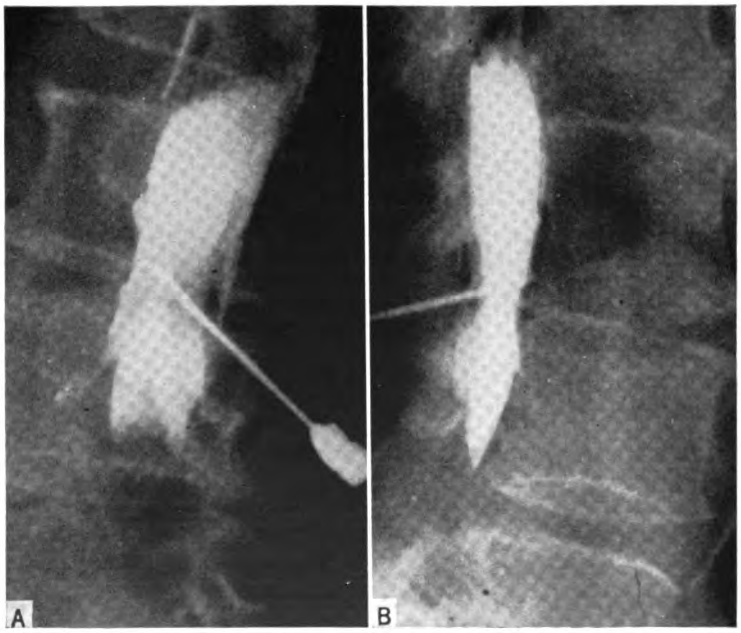

FIG. 156.—Complete spinal canal obstruction produced by a combination of a large ridge, heavy, thickened neural arch and ligamentum flavum, and a small discal herniation. Patient was a 65-year-old woman with low back pain radiating down both lower extremities, bladder dysfunction and weakness of both legs. Considerable relief after laminectomy and foraminotomy.

interspace, and the examination should be made in flexion and extension with the patient either sitting or erect. One should not overestimate the importance of the indentations caused by the infolding of the ligamenta flava during these maneuvers, especially in patients with relatively capacious canals.

Spur and ridge formations may produce clinical pictures indistinguishable from certain visceral diseases. Prominent among these is the simulation of anginal pain due to spondylosis of the upper dorsal spine. An excellent survey of this was made by Smith and Kountz (1942). On the basis of experimental movements made on a cadaver with spinal cord and nerves exposed they found that when the spine was either flexed or straightened to an abnormal degree the spinal canal tended to become elongated. The cord was drawn cephalad, thereby imposing tension on the spinal nerves at their angulations through the spinal foramina and at their attachments to the cord. It was postulated that this might give rise to tension and consequent irritation of the fibers producing referred pain simulating angina pectoris. Less well known is the syndrome of occipital pain. Josey (1940, 1951) suggested that such headache may be due to irritation of the muscle, fascia

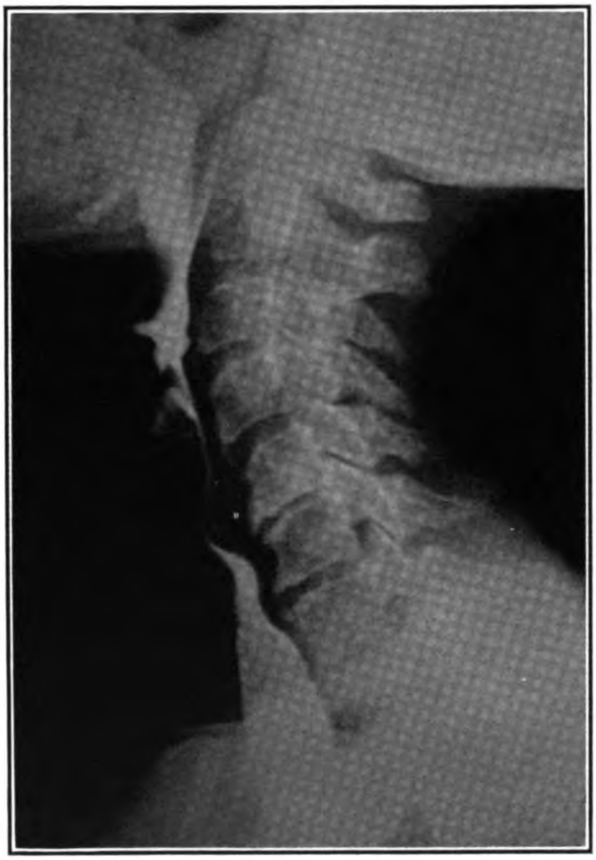

FIG. 157.—Lateral cervical esophagram in patient with asymptomatic cervical spondylosis. Note the indentation into the posterior aspect of the esophagus by the spurs at the fifth and sixth cervical interspaces.

and ligaments supplied by the upper three cervical and the ninth and tenth cranial nerves. Headaches of such origin may also extend forward to involve the temporal (Gunther and Kerr, 1929) and sometimes the frontal regions (Hartsock, 1940).

Mention has been made in various reports, particularly those of Gunsel (1952), Kertzner and Madden (1950) and Borak (1947), that large spurs in the anterior aspect of the cervical spine may be associated with dysphagia.

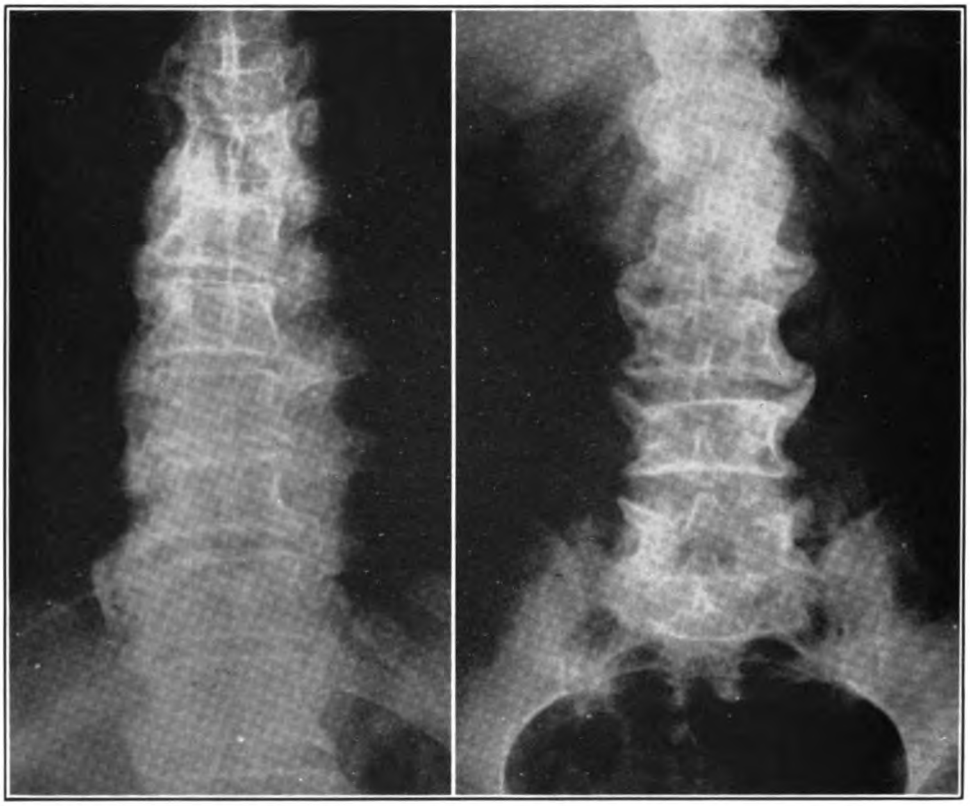

Fig. 158.—Anteroposterior roentgenograms of thoracic and lumbosacral spine with pronounced hypertrophic spondylosis in an elderly laborer with mild back pain.

The present author has found on routine esophagrams of patients with cervical spondylosis that defects in the posterior aspect of the barium-filled esophagus are present in many patients when the spurs project forward more than half a centimeter. This should not be accepted as a cause of dysphagia unless every other possibility has been excluded.

A form of spondylosis deformans worthy of comment is that associated with excessive bending and strain, such as may be seen in laborers or others engaged in lifting heavy weights. In these, large spurs appear at the lower thoracic and upper lumbar regions, and may be associated with considerable pain. With advancing age ossification of the paraspinal ligaments takes place as a physiological change, and often is without symptoms. This occurs more often in the thoracic spine, and is either limited to a few vertebræ or involves the major portion of this area. The ligamentous thickening varies from a few millimeters to as much as a centimeter in depth, and often appears to be separate from the vertebral margins. Increase in the dorsal convexity of the spine can be quite prominent. Relatively little spur formation occurs, but the intervertebral spaces become narrowed. The small joints may be relatively unaffected. The osseous densities are fairly well maintained unless concomitant osteoporosis is present.

With the more prominent changes of kyphosis senilis the alignment of the cervical vertebræ are altered so that an almost characteristic stance results. The patient maintains a posture in which the head is thrust forward so that the cervical spine is almost horizontally placed. In some patients these kyphotic changes are associated with emphysema and cardiac disturbances

Calcification of the posterior spinal ligaments is infrequent, but does occur occasionally and in the few cases I have seen the change was not associated with clinical manifestations.

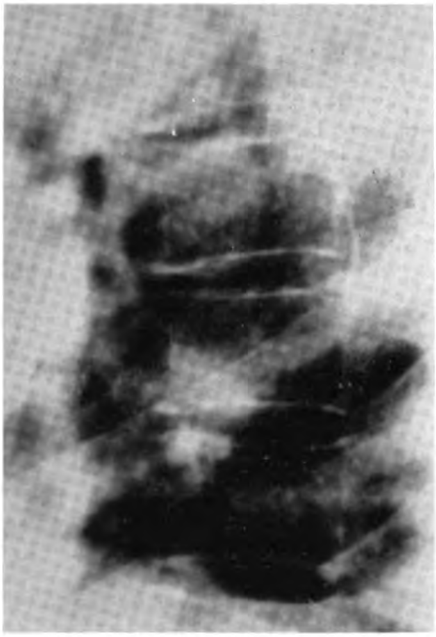

FIG. 159.—Calcification of posterior spinal ligament between sixth and seventh thoracic vertebra found on a lateral chest roentgenogram in a 45-year-old woman with no symptoms referable to the spine.

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HYPERTROPHIC OSTEOARTHROPATHY

Osteoarthropathy may occur as a concomitant of various cardiac and pulmonary diseases, especially chronic inflammatory diseases such as bronchiectasis and tuberculosis. It has also been observed with pulmonary tumors, liver and blood diseases and chronic intestinal conditions such as colitis. Clinically it may simulate rheumatoid arthritis (Temple and Jaspen, 1948). As a rule the condition is manifested mostly in the tubular bones and in the small bones of the hands. The predominant change is a roughening and thickening of the periosteum with consequent proliferation of the cortex. Involvement of the spine in hypertrophic osteoarthropathy is uncommon. In an extensive report of this condition Mendlowitz (1942) mentioned that the vertebræ were rarely involved.

Camp and Scanlan (1948) reported on several cases of a condition they termed chronic idiopathic hypertrophic osteoarthropathy. They defined this as a lesion occurring predominantly in males at the age of puberty or adolescence, and characterized by an osteoarthropathic syndrome of clubbing of the digits, enlargements of bones of the joints and thickening of the skin of the face in the absence of any primary disease. The disease manifests itself predominantly by thickening of subperiosteal bone following approximately the same distribution as noted in pulmonary hypertrophic osteoarthropathy. In their discussion they stated that the vertebræ were rarely involved.

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GOUT

Gouty involvement of the vertebral column has been reported rarely. An interesting case was described by Kersley, Mandel and Jeffrey (1950) in which an eighteen-year-old patient with gout was observed for five years. Death occurred because of sudden subluxation of the first cervical vertebra due to tophaceous softening, a condition not previously reported according to them. Lichtenstein, Scott and Levin described a vertebral body bone section as containing heavy urate deposits within most of the intervertebral discs and extending into the contiguous bodies. In the affected joints focal chalky white or yellow tophaceous deposits were present on the articular surfaces and bone ends as well as in the deeper layers of cartilage. Hall and Selin reported urate crystals deposits in the posterior articular surfaces of the fourth and fifth lumbar vertebræ, involving the ligaments and capsules as well. The crystals did not invade the cartilage deeply, but could not be separated even by firm wiping. This patient, a 51-year-old man, had had severe gout without back pain.

While there are no reports to the present authors' knowledge of specific spinal defects in gout other than those mentioned above, there appears to be clinical evidence that spine pain perhaps may be associated with this condition. In discussing symptomatology with many acquaintances with gout, I have the impression that even though the spinal *x*-ray examinations usually were negative, there was still reason for believing that gout may be associated with backache because of relief after appropriate medication. The pain described was of an aching, diffuse nature, and could not be sharply localized.

A case reported by Koskoff, Morris and Lubic (1953) is of particular interest. Their patient was a forty-four-year-old man with paraplegia due to extradural deposition of sodium urate crystals in the lower thoracic spinal canal. The plain films were normal, but myelography revealed a complete block at the level of the eleventh dorsal vertebra.

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INTERSPINAL OSTEOARTHROSIS ("KISSING SPINE")

An increase in the usual lordotic curve of the lumbar spine may produce approximation and contact of the tips of the spinous processes. This results in trauma and injury to the interspinous tissues and painful arthritic changes may follow. Other causes include Paget's disease, when the mass of the spinous processes are increased, or atrophy of the vertebral bodies and the intervertebral discs which results in approximation of the tips of the spinous processes. The condition may also be due to congenitally high spinous processes and low vertebral bodies or both anomalies together. It has been observed as part of the picture of spondylosis deformans. The lesion occurs most often in elderly patients. It is characterized by a chronic and sometimes severe pain, which may be aggravated by rotational movements or bending forward or backward. Radiologically, the tips of the involved spinous processes can be seen in contact with each other, and there may be bony sclerosis and overgrowth between the opposing edges.

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OSTEOMYELITIS

While pyogenic infection of the vertebral column is somewhat uncommon, it occasionally presents a perplexing problem in differential diagnosis. The usual cause is a staphylococcic metastasis from a distant focus of infection However, numerous other organisms may produce osteomyelitis of the spine. The association of vertebral osteomyelitis with genitourinary infections and after operations on the pelvic organs or the genitourinary tract has been attributed to spread of infection by way of the open system of veins along the vertebral column. The nutrient arteries of the vertebræ offer another and perhaps easier avenue of infection. A similar mechanism takes place in vertebral osteomyelitis complicating postpartum and other uterine infections. Osteitis pubis also represents a form of low grade infection which not infrequently follows prostatic surgery. Serial radiographic examinations are important in following the progress of the disease because of the lag between clinical and radiological manifestations. Body section radiography has been found to be of considerable help in evaluating small lesions. In patients with meningeal irritation or spinal canal obstruction myelography is essential, particularly in the presence of advancing neurological symptoms.

In pyogenic osteomyelitis of the spine the symptoms depend upon the site and the extent of infection into adjacent tissues. The complaints can be classified as predominantly cervical, thoracic, or abdominal and as involving the central nervous system, depending upon the extension of the bony involvement, and its relationship to the segmental nerves, pleura, and abdomen. The onset may be insidious or acute, and the progression of the infection varies considerably. A relatively benign form of spinal osteomyelitis is easily confused with tuberculosis. Associated with the bone infection are fever, leukocytosis, muscle spasm and signs of meningeal irritation when the infection extends into the spinal canal. With osteomyelitis of the lumbar spine the pain may be referred to the hip and leg, with pain, contractures, and limitation of motion.

The vertebral body is most often involved, but infection of the neural arches alone or even the spinous and transverse processes alone has been reported. The lumbar spine is the most commonly affected region, the dorsal spine is less commonly involved, and the cervical spine and coccyx presents the lowest incidence of osteomyelitis.

The radiologic evidences of osteomyelitis of the vertebral column usually lag considerably behind the pathologic changes, often as much as four to eight weeks. If adequate treatment is instituted, and the response satisfactory, no radiologic spine changes at all may develop. In the early stages of the disease involvement of bone may be limited to a small focus of rarefaction, usually in the superior or inferior aspects of the involved vertebral body close to the cartilaginous plate. Radiologic examination then may disclose nothing of significance, or at most a faintly decalcified focus. An early sign is slight narrowing of the intervertebral disc in the suspected area. When the disease has progressed so that there is appreciable destruction of bone, the rarefied area becomes more apparent. Collapse of the vertebral body may occur when the cancellous tissues are sufficiently weakened. Involvement of the intervertebral disc occurs frequently by direct extension through the cartilaginous plate. Adjacent to the intervertebral plate granular destruction of the bone takes place. When the infection has extended completely through the intervertebral disc to the supra- or infrajacent vertebral body, a roughly demarcated and rather ragged appearance of the opposing bone surfaces appears.

The disc itself is narrowed either by herniation of its contents into the vertebral spongiosa or by spread of infection into the disc. The degree of narrowing is variable. Osteomyelitis of the spine is frequently accompanied by rather rapid bony regeneration. Spur formations may appear at the same time as destructive changes. The appearance of proliferating bone in the presence of infection may be encountered within four to six weeks after the disease becomes apparent radiologically. The end-result of the disease

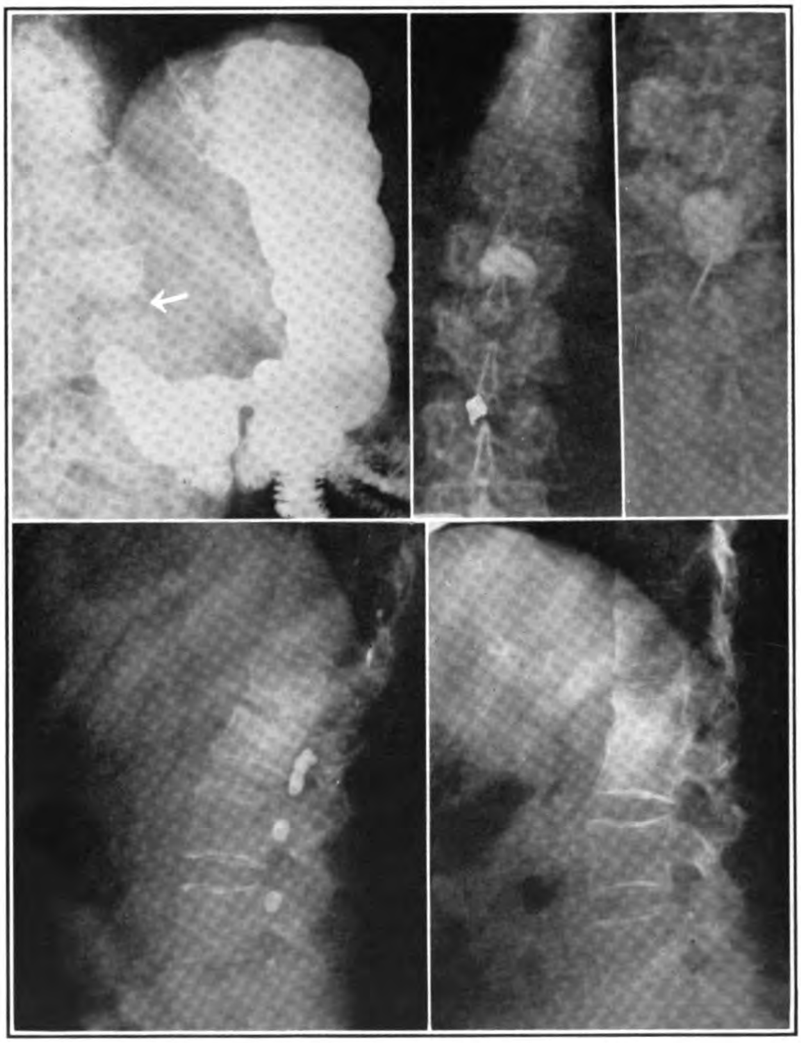

FIG. 160.—*A*, Right anterior oblique view of stomach. A destructive process is present at the first lumbar interspace, with involvement of the opposing bone surfaces. This was an incidental observation in a patient in whom the clinical diagnosis was perforating duodenal ulcer. *B*, Myelogram outlining the superior and inferior aspects of the intraspinal protrusion. *C*, Lateral spine roentgenogram made six weeks later, five weeks after operation when an inflammatory granulation was found impressing into the spinal canal. Some effort at bony repair is apparent. *D*, Seven months after identification of the lesion verified as osteomyelitis. Complete healing after antibiotic and chemotherapeutic treatment.

is a complete fusion of opposing vertebral bodies. Extension of the calcification outside of the confines of the vertebral bodies is infrequent, but occasional instances have been noted by us in which lime salts were deposited in abscesses adjacent to infected vertebræ. This tendency for osteomyelitis to heal with bone proliferation is considered useful in distinguishing osteomyelitis from tuberculosis of the spine.

With osteomyelitis of the spine several noteworthy complications can appear. Extension of infection from the cervical vertebræ into the soft tissues of the neck may result in marked widening of the retrotracheal space, with pressure against the dorsal aspect of the trachea and constriction of the esophagus. The usual lordotic curve of the cervical vertebræ is straightened, particularly when there is collapse of the anterior aspect of the involved vertebral segments. An interesting instance of osteomyelitis of the dens was reported by Frank (1944) in a forty-three-year-old man with a finger infection followed by cellulitis and bacteremia. *X*-ray examination of the neck at first was negative. Osteoporosis of the odontoid was noted within three weeks, followed by pathologic fracture through its base. Necropsy showed acute osteomyelitis of the odontoid process. Osteomyelitis of the transverse processes of the mid-cervical vertebræ was described by Finch (1947) in a baby three weeks old. This child was first seen with an abscess at the tip of a finger. When re-examined three weeks later, paralysis involving both arms, shoulders and hands was present. A lump appeared in the mid-line of the back of the neck, which became large and fluctuant. Thick green pus was aspirated, from which staphylococcus aureus was cultured. Healing took place by fusion of the involved vertebral bodies and their neural arches.

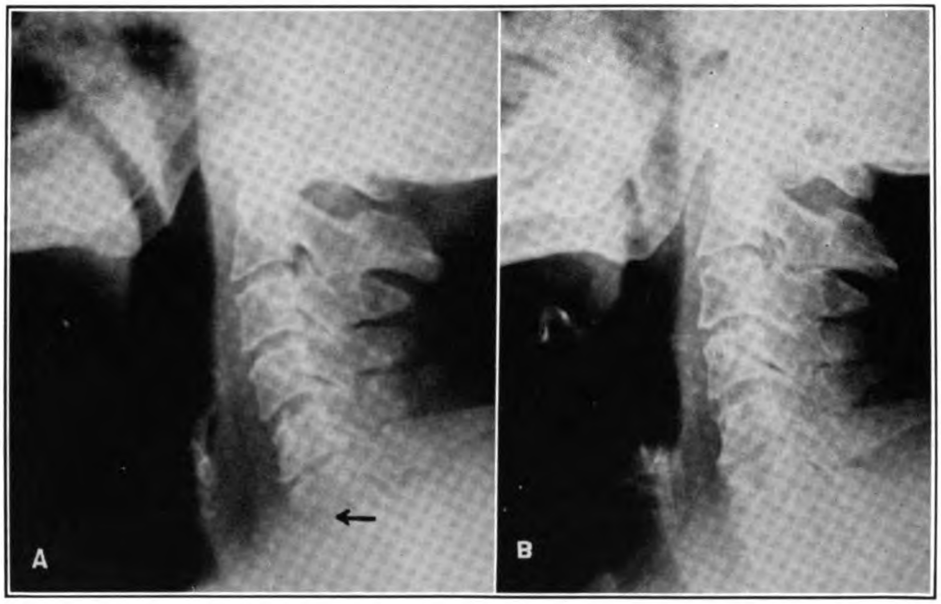

FIG. 161.—*A*, Osteomyelitis of the sixth and seventh cervical vertebræ, with destruction of the vertebræ anteriorly and widening of the retrotracheal space (arrows). *B*, Same case, six months later. Healing with conservative therapy. (This was in the days before potent specific therapy was available.)

Osteomyelitis of the thoracolumbar spine may invade the adjacent soft tissues and sometimes enters the thoracic cavity, perforating into a bronchus and manifesting itself by purulent expectoration. Such a case was described by Johnson and James (1945) in a thirty-four-year-old patient with suppurative osteomyelitis of the spine due to typhoid fever 22 years before admission. In this case a paravertebral abscess formed which perforated into the lung. Brill and Silberman (1938) described a case of pyogenic osteomyelitis of the spine with mediastinal abscess and spinal cord compression.

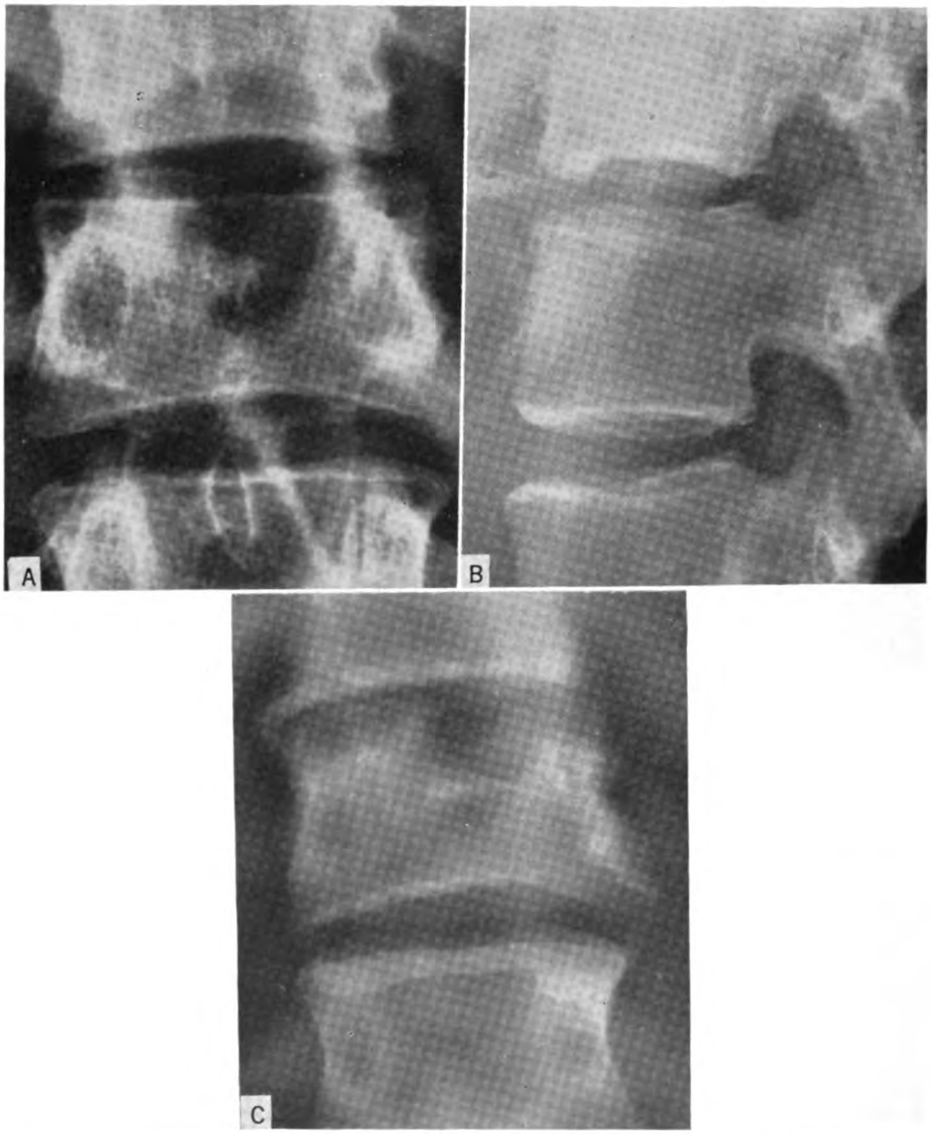

FIG. 162.—Osteomyelitis of first lumbar vertebra. A loss of continuity of the anterior portion of the superior surface is seen on the anteroposterior film (*A*). The lateral film (*B*) shows a slight irregular loss of bone of the anterosuperior aspect of the vertebral body. The body section roentgenogram (*C*) clearly demonstrates the lytic lesion in the superior aspect of the vertebral body. Note that the intervertebral disc is not grossly disturbed in this patient.

Extension of osteomyelitis into the soft tissues adjacent to the lumbar vertebræ and psoas abscess formation is not infrequent. Sinus formation may occur with extension of the infection to more remote areas, sometimes emerging to the skin. In such case injecton of the fistuous tract with lipiodol or water-soluble contrast materials may help identify the source of the infection. We have observed several cases in which osteomyelitis of a lumbar vertebra was limited to a transverse process, and radiologically presented only a small area of rarefaction. The contrast injection in these people disclosed large sinus tracts with purulent collections deep in the tissues adjacent to the spine, and extending a considerable distance from the original small infected area. Shehadi (1939) reported a case of osteomyelitis involving the inferior articular process of the second lumbar vertebra, in which serial roentgenograms over an eight-year period revealed a gradual disappearance of this structure. Cure was spontaneous.

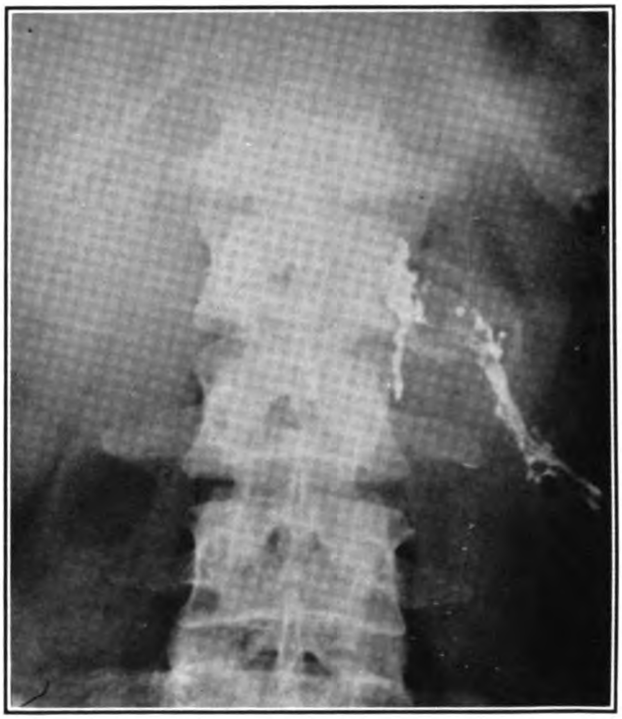

FIG. 163.—Lipiodol injection of sinus tract leading to the primary osteomyelitic focus in the left transverse process of the second lumbar vertebra.

Osteomyelitis of the neural arches is not as frequent as involvement of the vertebral bodies. It may be associated with extension of the infection into the epidural space, producing spinal cord irritation and compression. In such cases myelography may disclose evidence of block and constitutes an important part of the examination.

Instances of infection of vertebral bodies after trauma are infrequent. Such a case was reported by Atstatt (1939) in which chills and fevers followed fracture of the second lumbar vertebra due to a fall. This patient developed a secondary hematogenous osteomyelitis secondary to a streptococcus infection, as disclosed by necropsy.

The role of lumbar puncture in the development of osteomyelitis of the spine has been recognized. Redo (1953) reported 4 cases of vertebral osteomyelitis following lumbar puncture, bringing to a total of 57 the number of these cases. Most were associated with a difficult spinal tap. Roentgenograms revealed narrowing of the involved intervertebral disc, followed by sclerosis or rarefaction and destruction of the involved vertebral bodies.

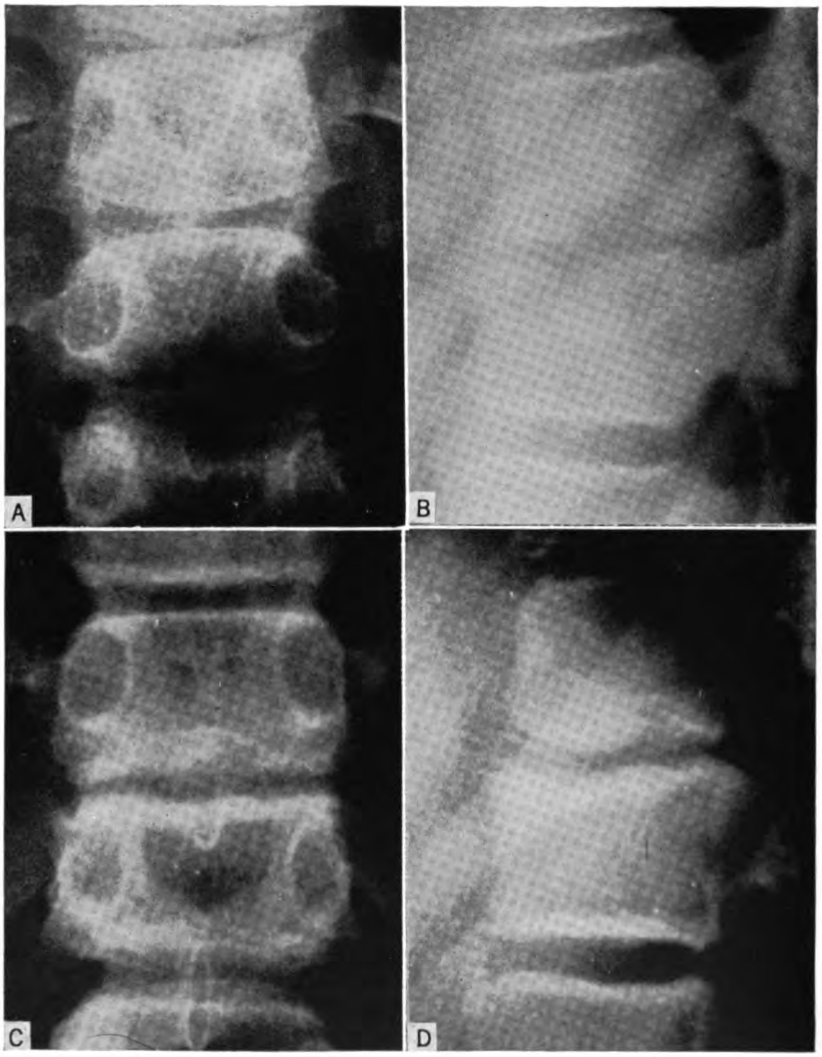

FIG. 164.—Osteomyelitis involving the tenth and eleventh thoracic vertebræ in a 9-year-old girl. *A* and *B*, Anteroposterior and lateral roentgenograms reveal demineralization of the involved bony surfaces and narrowing of the intervertebral space. *C* and *D*, Four months later, conservative treatment followed by healing. Note the sclerotic changes. The narrowed interspace persists.

The occurrence of osteomyelitis of the lumbar spine following operation for herniated disc has been observed by us in 4 instances. In all, surgery had been preceded by myelography. However, the infection first manifested itself in the anterior aspect of the involved vertebral bodies, and infection appeared clinically about two weeks after operation had been done. In 2 of these cases a slow but satisfactory response followed antibiotic therapy. In the other the response was not as satisfactory, and the infection took a long time before healing by sclerosis took place.

Osteomyelitis of the sacrum and sacroiliac joints may be directly introduced, follow infections in the adjacent soft tissues or be of metastatic

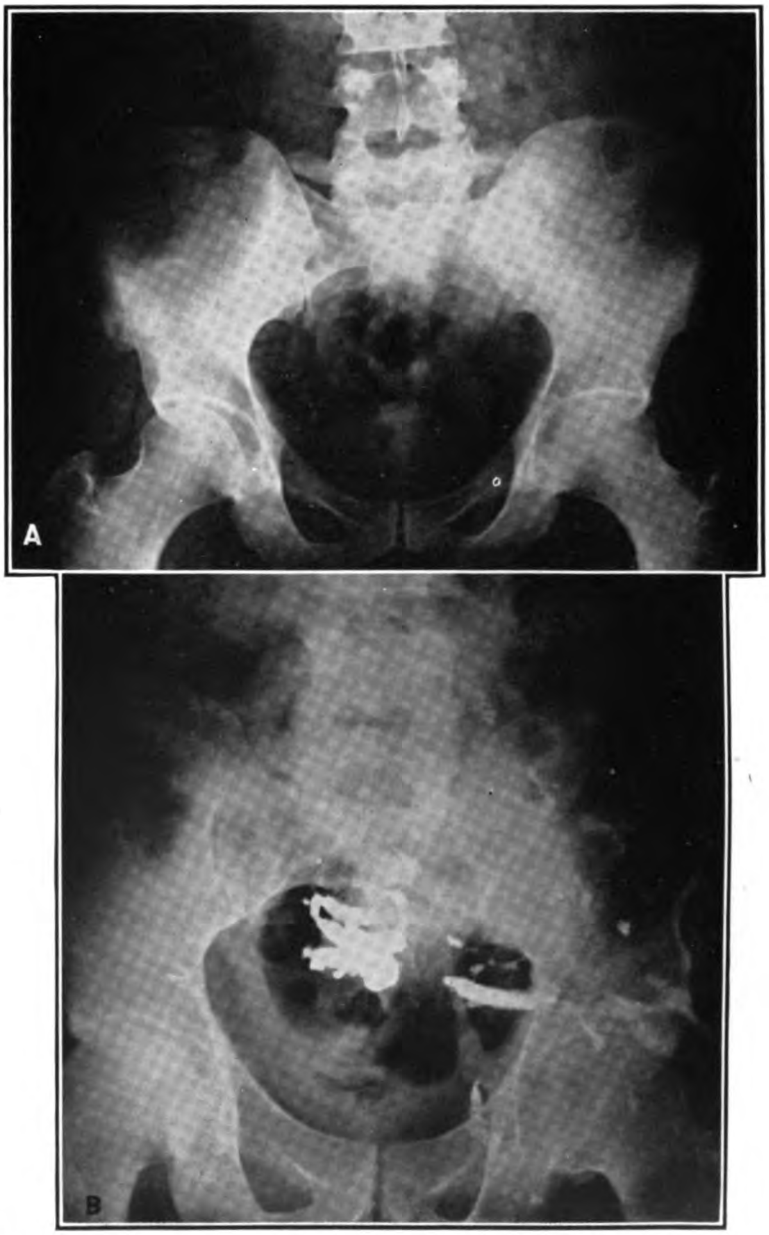

FIG. 165.—*A*, Osteomyelitis of the body of the sacrum. A sinus tract extends to the left hip region. A rarefied destructive focus is present in the sacrum, overshadowed by gas in the bowel. *B*, Same case, lipiodol injection of the sinus tract. The contrast medium collects in the hollow of the sacrum.

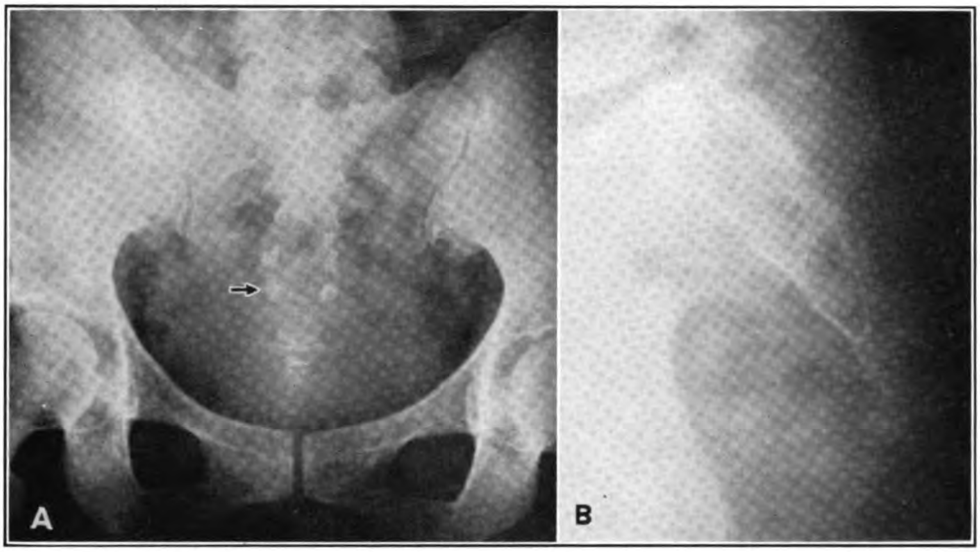

FIG. 166.—*A*, Osteomyelitis of lower sacrum in a patient with lymphogranuloma venereum. Note the destruction of the pedicles of the midsacral segments (arrows), and the adjacent bone. *B*, Same case. The anterior aspect of the lower sacrum is eroded.

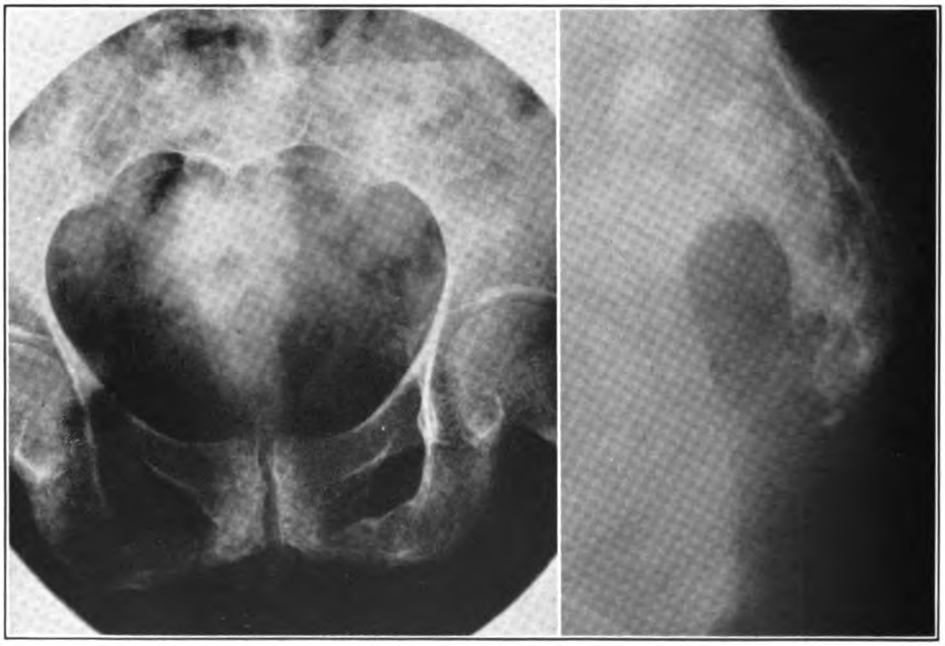

FIG. 167.—Healed osteomyelitis of the lower sacrum, with heavy bone deposition.

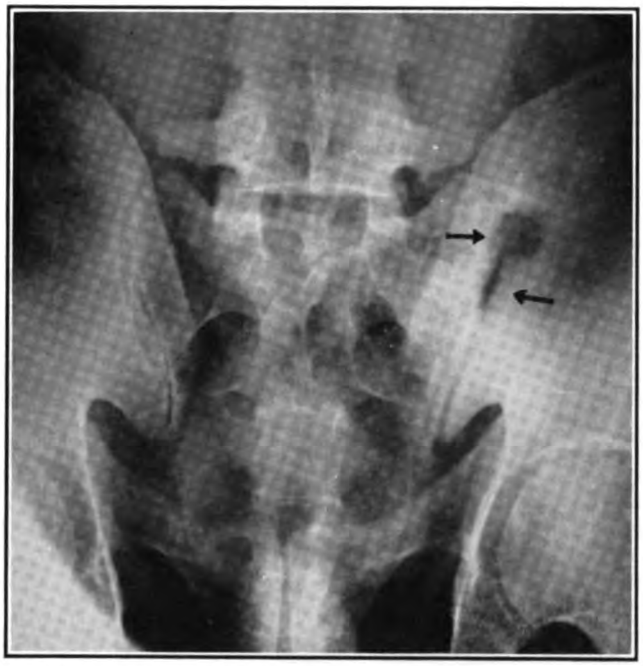

FIG. 168.—Osteomyelitis of the left sacroiliac joint. Sclerotic changes, with an elongated irregular oval area of destruction in the upper half of the joint (arrows).

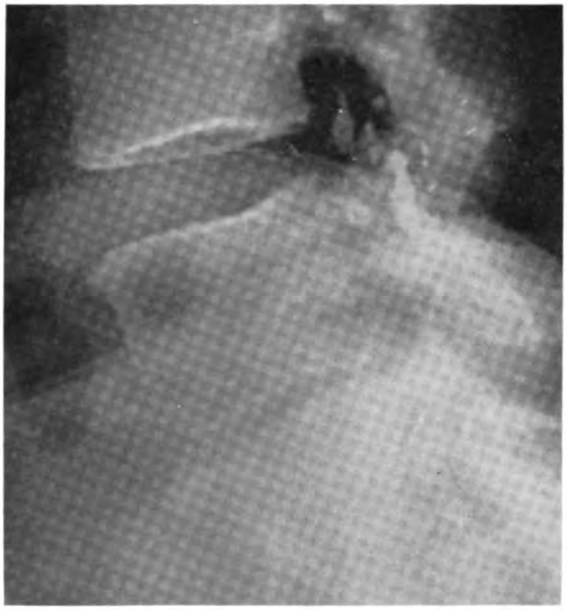

FIG. 169.—Lateral roentgenogram of lumbosacral spine. Osteomyelitis destroying the antero-inferior aspect of the fifth lumbar vertebra following operation for herniated disc.

origin. The infection may involve the body of the sacrum, with destruction of its midportion and formation of sinus tracts to its immediate vicinity or extending as far distally as the hip region. With healing, the lytic areas gradually fill in and eventually become densely sclerotic. The process may be indolent and last a long time, resulting in destruction and deformity of the distal sacrum. In the 3 cases illustrated here 2 followed caudal anesthesia for obstetric delivery, and 1 was associated with lymphogranuloma venereum. Another case followed metastatic extension from a skin infection.

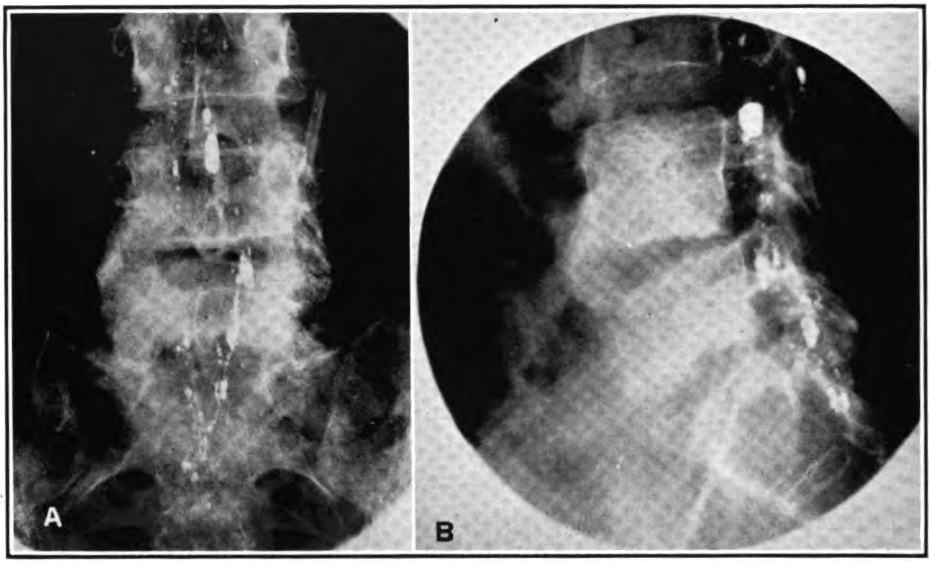

FIG. 170.—Osteomyelitis following operation for herniated disc, with bony repair. The anterior aspects of the fourth and fifth lumbar vertebræ are involved.

A benign form of osteitis of the spine in children was reported by Bremner and Neligan (1953). This appeared with symptoms of a systemic infection in children from nine months to two-and-a-half years of age in his series, followed by pain in the back and stiffness which prevented handling, sitting or walking. The symptoms progressed gradually for about five or six weeks, then became stationary or receded. Narrowing of the intervertebral discs, erosion of the adjacent vertebral margins, and sometimes lateral or posterior displacement of an involved vertebra occurred. Healing was manifested by recalcification of bone. The causative agent was believed to be staphylococcic.

Rarely osteomyelitis may present as a densely sclerotic vertebra, simulating a so-called "ivory vertebra." We have had but one verified example of this unusual variation in the roentgenologic picture of osteomyelitis in a 16 year old boy who responded quite well to treatment by prolonged rest and antibiotic medication. The density of the vertebra did not alter perceptibly over a period of observation of about a year, even though symptoms disappeared. This patient is still under our care.

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Typhoid Osteomyelitis

Typhoid osteomyelitis may develop long after the infection supposedly has been cured. Spota and Bardeci (1950) reported a forty-four-year-old woman with osteomyelitis of the upper dorsal spine twenty years after typhoid infection. The neural arch was involved to a greater extent than the body, and paraplegia developed because of extension of infection into the spinal canal. Subarachnoid block was due to extradural pressure from a typhoid abscess. Steingraber (1947) mentioned that typhoid spondylitis may appear during the convalescent period, approximately four weeks after the disappearance of symptoms.

The basic pathologic change is bone destruction caused by infected embolic infarcts. The intervertebral disc is secondarily affected by rupture of the focus through the cartilage, much the same as in pyogenic osteomyelitis (Keith and Keith, 1926). Healing is associated with osteosclerosis and bone bridging, and may take more than a year to complete.

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Brucella Spondylitis

Involvement of the spine with undulant fever is frequent, varying according to different authors from 10 to 65 per cent. Males are affected much more frequently. The disease affects the vertebral bodies, the neural arches, and extends into the intervertebral discs and the adjacent ligaments. Further progression into the spinal canal, with involvement of the meninges and the adjacent nerve roots may take place. The condition may be extremely painful and produce bony changes in multiple and sometimes separated vertebræ (Purriel, Risso and Espasandin, 1943). From the radiologic viewpoint, DiRienzo (1943, 1950) reported that an early sign was lack of definition of the bone structures, with some attempt at bone regeneration which may appear in the more chronic forms. Regeneration of bone sometimes is quite marked. Extensive bony overgrowth may be concomitant with newly formed destructive areas. In young people the destruction of the vertebral bodies and the intervertebral discs may be extensive. At the end stages of the disease the appearance is that of a chronic destructive spondylitis. Calcification of the anterior spinal ligament is fairly common, that of the posterior spinal ligament is rare.

Spink (1948) reported that infections with Brucella abortus in Minnesota was often associated with bone pathology. He presented an instance of osteomyelitis of the cervical spine, acute in nature, with partial collapse of the inferior aspect of the third cervical vertebra which healed within a month with new bone formation. Bishop (1939) had mentioned that vertebral lesions in undulant fever were characterized by increased density of the involved vertebræ, preservation of the body contour and relatively minor changes in the intervertebral disc together with hypertrophic spurring. According to him there was little tendency to invasion of the paravertebral tissues.

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SYPHILIS OF THE SPINE

Congenital syphilis is due to infection of the fetus through the placental circulation. Destructive changes take place, particularly in the growing ends of the extremities, with granulation tissue replacing bone. Periosteal elevation, osteoporosis and nonspecific band-like zones of radiolucency also appear. Osseous changes in the vertebræ with congenital syphilis are rare. McLean (1931) reported zones of increased and diminished density in the superior and inferior aspects of involved vertebræ, with no visible bone destruction. He considered these to correspond with the transverse bands present in the long bones. Péhu and Boucomont (1933) reported a fifteen-month-old infant with syphilitic spondylitis involving the twelfth thoracic and first and second lumbar vertebræ. A gibbus was present, but no paravertebral swelling was seen. Tissue removed from this region was described as being consistent with but not pathognomonic of syphilitic spondylitis.

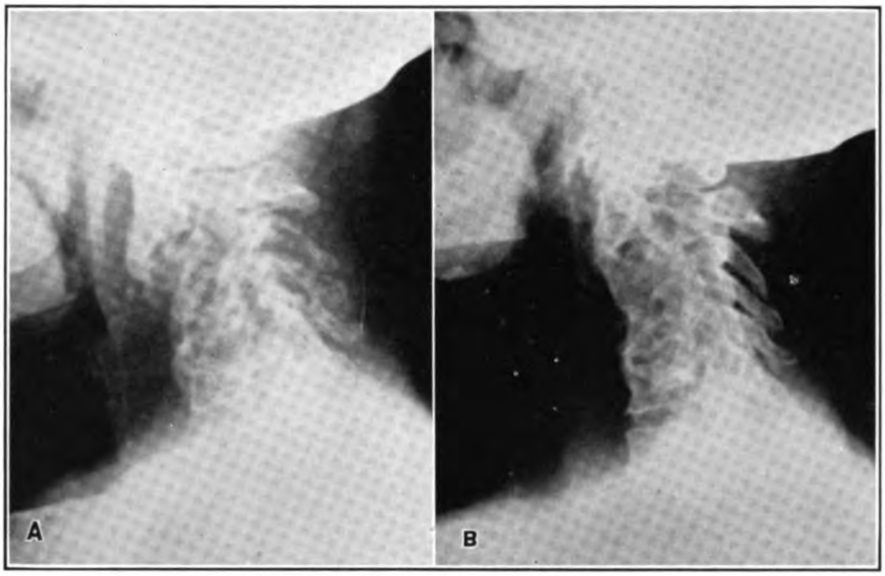

FIG. 171.—*A*, Syphilitic osteitis of the cervical spine, with involvement of the bodies, pedicles and spinous processes. Irregular, diffuse bone destruction, with loss of usual cervical curve. *B*, Same case, three months after antisyphilitic treatment (1933). Note the repair of bone, with fusion of the vertebral bodies and the regeneration of the spinous processes.

Vertebral syphilis in later life follows periosteal and osseous spirochetal invasion, producing osteoperiostitis of the cortex and medulla of the involved vertebræ, with granulomatous extension into the adjacent soft tissues. Occlusion of blood vessels by syphilitic arteritis causes bone destruction. The irregular foci of destruction and reconstruction of bone are usually more marked in the anterior portions of the involved vertebræ. The intervertebral discs are quite resistant, but with extensive involvement may become narrowed or almost completely destroyed.

The cervical vertebræ are involved most often with syphilitic spondylitis. Three such cases were reported by Sgalitzer (1941), with intense pain and destructive lesions involving particularly the ventral aspects of the involved bodies. The dorsal spine is not affected as often as either the cervical or the lumbar vertebræ.

The radiologic appearance of syphilis of the spinal column varies greatly. When only a single vertebra is involved the diagnosis may be most difficult to make, the only evidence being a destructive process in the anterior aspect of the vertebral body. When reparative changes take place the presence of some overgrowth of bone into the paravertebral soft tissues may offer some diagnostic help. With more extensive involvement the concomitant destruction and production of bone and the overflow of reparative bone into the adjacent soft tissues is more easily recognized. The intervertebral spaces may be visible even though extensive destruction occurs adjacent to them, while close by irregular deposits of ossification are prominent. Bridging between the involved vertebræ may be quite extensive, and the calcification may involve the paraspinal ligaments.

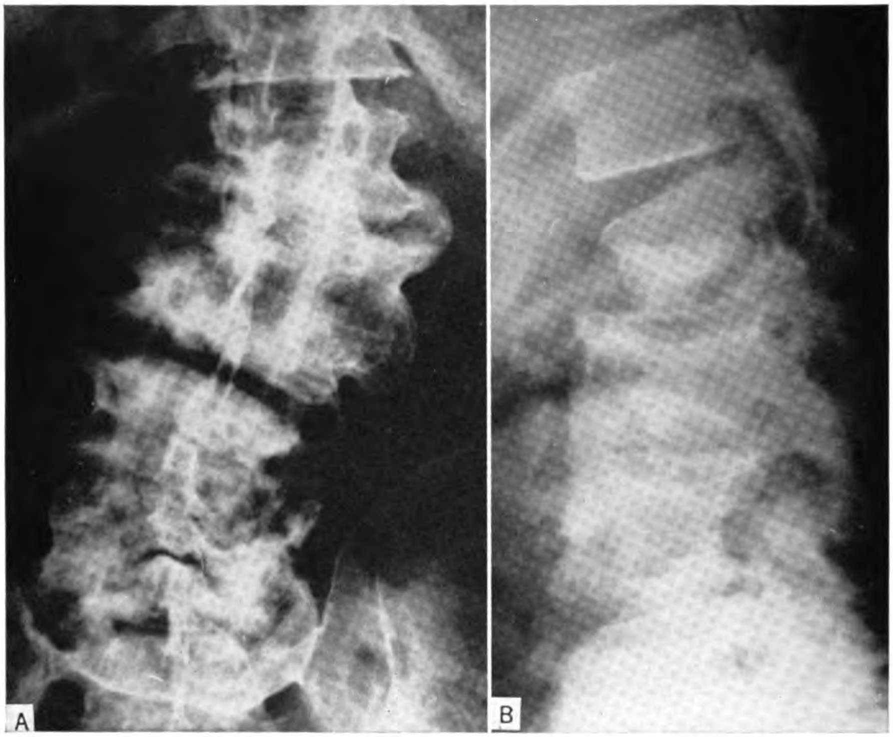

Fig. 172.—Syphilitic osteitis with heavy bony repair, large osteophytes and shifting of the vertebral bodies.

In our own experience there were 5 cases of syphilitic spondylitis in adults. Two had extensive destruction of the midcervical vertebræ, with marked distortion of the vertebral bodies and practically absent intervertebral discs. In both the cervical spine remained almost vertical in alignment, probably because healing after specific treatment had rendered the midcervical spine a practically solid, irregular column of bone. The other cases presented involvement of the second, third and fourth lumbar vertebræ. The involved bodies were dense and sclerotic. The intervertebral space between the second and third lumbar vertebræ was narrowed and irregular, but still could be identified. That between the third and fourth lumbar vertebræ was narrowed posteriorly to a considerable extent, but open and smooth anteriorly. Heavy solid perivertebral calcification with large spurs was present, more pronounced anteriorly and to the left side. The heights of the second and third lumbar vertebræ were reduced by about half, while the fourth lumbar vertebra was only slightly decreased. Because of the irregular destruction and heavy bony reaction of the opposing surfaces of the second and third lumbar vertebræ, they appeared flattened and broadened. In addition, lateral displacement due to change in alignment could be seen (Fig. 172).

The occurrence of gummas within the spinal canal is rare. Such a case was reported by Ray (1940) in a forty-one-year-old male who presented a clinical syndrome consistent with the diagnosis of a tumor of the cauda equina. At operation the nerve roots were slightly swollen and loosely stuck together by what appeared to be a low-grade inflammatory process. A yellowish-white tumor mass about 2 cm. in its widest diameter was encountered. The tumor appeared to rise in one of the main roots on the left side, and was slightly adherent to the dura. Because of the general inflammatory appearance of the region, a syphiloma or tuberculoma was suspected. The diagnosis of syphilis was confirmed by examination of a biopsy specimen. His patient did quite well on anti-syphilitic treatment.

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NEUROPATHIC JOINTS (Charcot's Arthropathy)

Included in this group are cases with changes in the spine usually secondary to central nervous system syphilis. Rarely it may be observed in patients with diabetes, cauda equina tumors, spina bifida, traumatic paraplegia, transverse myelitis, and hematomyelia (Holland, 1952). The primary pathologic process occurring in a neuropathic joint is one of degeneration and partial disappearance of the articular cartilages which are invaded by fibrous connective tissue from an inflammatory pannus of its surface. The zone of preliminary calcification becomes exposed by stripping away of the cartilage, beneath which further cartilage may proliferate and become converted into subchondral bone. This accounts for the sclerotic eburnated bone seen at the base of large defects in the articular cartilages. A generalized atrophy of the trabeculæ of the cancellous bone is often seen. As a result of this combined degeneration of cartilage and proliferation of subchondral bone, osteophytes develop either as free bone fragments or as amorphous dense scattered conglomerates which appear to grow in a purposeless fashion. Such bony proliferation in the spine occurs first in the regions of the articular margins, resembling hypertrophic spondylosis. Associated with this are areas of degeneration of bone resulting in radiolucent foci within malformed vertebral bodies. The appearance of bone atrophy and degeneration, together with the extensive deposition of proliferating bone both adjacent to the vertebral column and some distance from it is fairly characteristic of the Charcot joint. Associated with these changes may be calcification in the adjacent soft tissues, particularly in the muscles and ligaments.

The usual site of vertebral osteoarthropathy is the lumbar spine. Occasionally the dorsal vertebræ or a single vertebra may be involved. The differential diagnosis between neuropathic osteoarthropathy and syphilitic osteoperiostitis of the spine on purely radiologic grounds may be difficult. The more frequent involvement of the cervical spine with syphilitic spondylitis, and the purposeless diffuse proliferation of bony growth into the soft tissues seen with Charcot's disease may be of some help. Incidentally, the blood and cerebrospinal fluid Wassermann reactions may be normal in about half the cases.

The cause for neuropathic disturbances in the joints is commonly ascribed to trauma, combined with analgesia and consequent loss of warning pain. In an analysis of 134 cases, with a total of 214 Charcot joints, Steindler, Williams, and Puig-Guri (1942) noted that in only 57 was there a definite picture of tabes. In discussing pathogenesis, Delano (1946) noted that the explanation of all neuropathic arthropathies was ascribed to a common origin of trophic disturbance. He regarded this as inadequate to explain the lavish and purposeless production of new bone around the involved joints which to him suggested that a profound disturbance of regional nutrition was taking place. In his report he describes a patient paralyzed from the waist down for fifteen years following a fracture of the first lumbar vertebra. Radiographic examination showed extensive irregular bony overgrowth and osteosclerosis, consistent with joint neuropathy.

The occurrence of other neuropathic joints in association with Charcot's disease of the spine is well known. Such a case was recently described by Goodman (1949) in a sixty-four-year-old white male with tabetic neurosyphilis in which the elbows, knees and hands were involved as well as the lumbar spine. Neuropathic joints associated with syringomyelia are reported most often in the upper extremities, but sometimes involve the lower extremities (Skall-Jensen, 1952). Involvement of the spine, if it occurs at all, is most unusual. I have not seen vertebral neuropathic disease caused by syringomyelia.

The association of diabetes with neuropathic joints has come into considerable prominence lately. Knuttson (1951) reported 4 cases involving the joints of the lower extremities, but no involvement of the spine was mentioned. Four additional cases with osseous destruction of the feet were reported by Beidelman and Duncan (1952). No spinal involvement was noted by these authors. Zucker and Marder (1952) presented a case of Charcot spine due to diabetic neuropathy in a patient with severe pseudotabes. They believed this to be the first case of Charcot spine reported as a result of diabetic neuropathy. Extensive destruction and disorganization of the second, third and fourth lumbar vertebræ were present, together with considerable bone production within the area of the destroyed vertebræ. The etiologic and pathologic aspects of this patient were confirmed at necropsy.

We observed a male patient forty-five years old with Charcot's disease of the spine (Fig. 173) who presented dissolution of the superior half of the body of the second lumbar vertebra, so that the intervertebral space between the first and second lumbar vertebræ appeared relatively widened. Large, heavy, dense bony bridges between the vertebræ apparently were strong enough to keep them apart. Large amorphous calcific deposits were spread in the adjacent soft tissues for more than 3.5 cm. to either side as well as anteriorly. The anterior aspect of the first lumbar vertebra inferiorly and the second lumbar vertebra superiorly were partially destroyed, and a slight gibbus formation was present. Heavy calcification of the spinal ligaments was present. This patient suddenly became paraplegic, and myelography revealed a block at the level of the first lumbar vertebra. At operation this was found to be due to compression of the spinal canal by granulomatous tissue from the involved bony walls.

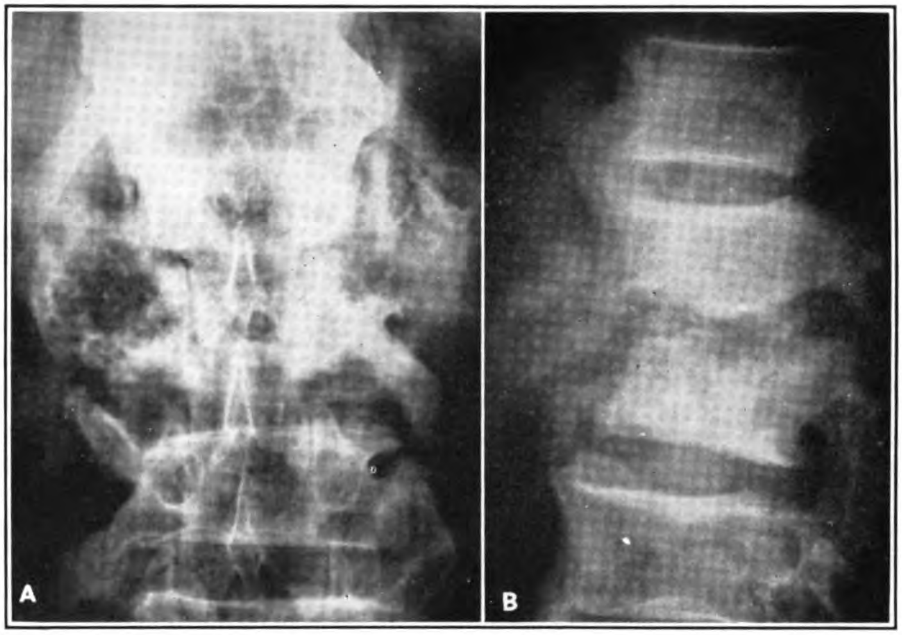

FIG. 173.—Charcot's disease of the upper lumbar vertebræ, with prominent bone destruction, large amorphous calcific deposits adjacent to the involved first lumbar interspace.

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FUNGUS, PROTOZOAN AND PARASITIC INFESTATIONS

Actinomycosis follows infection with fungi of the actinomyces group, the most frequent being actinomyces bovis. Other members of the genus may also be pathogenic, but the clinical manifestations are identical. The portal of entry is usually the mouth, the organisms resting in carious teeth, pyorrhetic gums or the tonsils. They assume pathogenicity upon entrance into the subcutaneous tissues after local trauma or other disease processes. The characteristic granulomatous lesions produce local swellings in the jaws, the so-called "lumpy jaw." Abscess formation is accompanied later by sinus tracts discharging to the skin, and from this pus may be cultured the causative fungus, or the characteristic sulfur granules may be identified as yellowish-gray granular masses.

Actinomycosis can then spread to other regions, particularly the chest and the gastrointestinal tract. Vertebral involvement may follow extension of the granulomas to the paravertebral soft tissues. Less often infected emboli rest in the medulla of a vertebra and set up an intraosseous granuloma. When the outside tissues are infected extension to the periosteum and the cortex results in destruction of bone. A reparative effort is manifested by the appearance of bone sclerosis which may be quite intense at the margins of the infected bone. With extension of the disease numerous channel-like areas of destruction permeate the vertebra producing an appearance variously described as "soap-bubble," "honeycomb" or "lattice-like." This may involve all or any part of the vertebra including the spinous and transverse processes. From this, infection extends to the adjacent ribs, visceral cavities or the skin with multiple sinus tracts. As a rule, multiple vertebræ are diseased, but it is not uncommon in this relatively rare condition to observe involvement of a single vertebra, usually depending on the point of origin of the infection. The intervertebral discs as a rule are spared, and collapse of involved vertebræ even when extensively diseased is uncommon, although occasional instances of angular deformities are known (Lubert, 1944).

The symptoms of vertebral actinomycosis are inconstant and varied. Pain, tenderness, limitation of motion and nerve root pain are all dependent on the location of the disease and its spread to adjacent soft tissues. Prior to the advent of chemotherapeutic and antibiotic agents the prognosis was poor, but this has now changed for the better (Brett, 1951; Cope, 1951).

Involvement of the spinal canal with actinomycosis is rare. Such a case with meningeal involvement and compression of the spinal cord was reported by Lamartine de Assis and Mignone (1946) in a fifty-two-year-old male with cutaneous and thoracic spine lesions. Necropsy disclosed purulent exudate in the tissues around the spinal column, and there was extensive thickening of the dura with spots of inflammatory exudate. The cord was intact, and was not actually involved.

Blastomycosis (American blastomycosis; Gilchrist's disease) is caused by infection with the fungus Blastomyces dermatitidis. The portal of entry is the skin, upon which small papulo-pustules form. These gradually enlarge, become crusted over and form irregular, patchy papillomatous tumors which expand peripherally and heal centrally with a flat scar. This local form of blastomycosis is relatively benign. When systemic dissemination occurs there may be involvement of the lungs, the internal organs and the bones. Occasionally the primary focus is in the lungs, and the lesion may be mistaken for tuberculosis or bronchogenic carcinoma.

Meyer and Gall (1935) mentioned 12 cases of blastomycosis of the spinal column in a review of mycotic diseases of the spine. In a review of 63 cases collected from the literature by Jones and Martin (1941), 25 instances of vertebral involvement were noted, and paravertebral abscesses were observed in 14. Colonna and Gucker (1944) observed that the bony manifestations of blastomycosis suggested an ordinary osteomyelitis and presented no characteristic features. Baylin and Wear (1953) indicated that spinal blastomycosis might simulate tuberculosis.

Involvement of the spinal canal is rare. Craig, Dockerty and Harrington (1940) reported a case of an intravertebral and intrathoracic blastomycoma which simulated a dumbbell tumor. Another case with spinal cord involvement was reported by Greenwood and Voris (1950), with erosion of the pedicles of the sixth and seventh dorsal vertebræ and foci of destruction in the body of the eighth dorsal vertebra.

Coccidiomycosis ("valley fever") is caused by infection with the fungus Oïdium coccidioides (Coccidioides immitis). The portal of entry in most cases is the pulmonary tract, and the initial lesions resemble tuberculosis, with a clinical picture resembling an upper respiratory infection. Recovery from the primary infection usually takes place within a few weeks, leaving the patient hypersensitive to the organism. Re-infection endogenously or through the skin may introduce the secondary or granulomatous phase, and the disease enters a prolonged period of exacerbation and remission during which time metastatic involvement of the skeleton as well as the visceral organs may occur. The disease occurs more often in males, and only a small proportion of infected patients develop the granulomatous phase. According to Benninghoven and Miller (1942) the lesion may exist in the spine as a single central focus, but most often multiple marginal vertebral changes take place because of extension to the bone from paravertebral abscesses. Miller and Birsner (1949) stated that in their series of 17 cases there was no bone involvement caused by extension from soft tissue abscesses or granulomas. The spine was involved in 6, most frequently the lumbar vertebræ. The lesion noted by them was predominantly lytic, with no involvement of the intervertebral discs. Some sclerosis was present, and they noted linear densities which they considered calcification in the paravertebral soft tissues. Denenholtz and Cheney (1944) reported that in a case observed by them eight and one-half months after the onset of the disease there was destruction of the intervertebral disc between the tenth and eleventh dorsal vertebræ as well as osseous erosion. They suggested that differential diagnosis from tuberculosis might be quite difficult.

Torulosis (*Cryptococcosis; European blastomycosis*) is a fungus infection due to the Torula histolytica (Cryptococcus hominis). The portal of entry is believed to be the upper respiratory tract from which there followed disseminated discrete lesions. In a summary of 10 cases from the literature, including 1 of their own, Carlton and Mount (1951) discussed the neurosurgical aspects of torulosis. In their case a cauda equina tumor was simulated. A spinal tap disclosed cloudy fluid with a total protein of 4200 milligrams per cent. The plain spine roentgenograms were normal. At operation a laminectomy from the first to fourth lumbar vertebræ disclosed a solid, firm, tenacious gray-white mass occypying almost the entire spinal canal from the upper portion of the first lumbar vertebra to the upper portion of the fourth lumbar vertebra. The arachnoid was thickened, and when this was opened, 2 to 3 cc. of purulent fluid exuded, from which the Crypotcoccus neoformans was recovered. Ley, Jacas and Oliveras (1951) recorded a case of torula granuloma of the cervical spinal cord in which the plain films showed widening of the spinal canal and erosion of the pedicles. This occurred in an eight-year-old child with quadriparesis. The true nature of the disease was not known until after histologic study showed the characteristic torula organisms. The lesion seen at operation was an extensive granuloma simulating a spinal cord tumor.

Bone involvement in cryptococcosis was discussed by Collins (1950) who mentioned that in over 200 reported cases 17 examples of bone involvement were encountered. In his first case a destructive lesion was present in the upper three lumbar vertebræ, with cortical erosion and no osteoblastic reaction in the soft tissues. Abscess formation in the adjacent lumbar musculature was noted. In his second case, abscesses were present in the cervical lumbar and sacral regions with areas of bone destruction in the pedicle of the fourth lumbar vertebra.

It is not possible to distinguish one fungus infection from another radiologically.

Toxoplasmosis

This is a disease caused by toxoplasma, a protozoan organism of uncertain status, usually classed with the sporozoa. This protozoan may infect many tissues, the central nervous system being often involved. It was first described as causing an encephalomyelitis in infants, giving rise to a syndrome of hydrocephalus accompanied by convulsive seizures, enlargement of the head and increased intracranial pressure. Radiographic examination of the skull often shows calcifications in the cerebral hemispheres. In a report by Dyke, Wolf, Cowen, Paige and Caffey (1942) mentioned that in the vertebral column the provisional zones of calcification in the superior and inferior margins of the vertebral bodies were exaggerated into heavy white borders. Internal to the heavy white lines were parallel linear zones of decreased density similar to the deep transverse zones of the ends of the shafts of the long bones. These authors considered the lesions as probably not specific for toxoplasmosis since they had been observed in several other infectious and nutritional disturbances.

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PARASITIC INFESTATIONS OF THE SPINE

Echinococcosis (Hydatid disease).—This condition results from the ingestion of the ova of echinococcus granulosus. Infestation is usually incurred during childhood from contact with dogs or cats. Unless there is involvement of the brain or the orbit, symptoms do not develop until later in life when the slowly growing tumors make themselves apparent. Involvement of the spine with hydatid disease has been described by many authors. Bellini (1946) reported that bone involvement occurred in approximately 1 per cent of cases. The six-hooked embryo reaches the bone by way of the arterial circulation, and establishes itself in highly vascularized areas such as the epiphyseal ends of long bones or the spongy bone of the vertebral bodies. Vertebral hydatosis shows no tendency to formation of cyst-like spaces, and for that reason the height of the vertebræ usually is not changed and the intervertebral cartilages remain undisturbed. Often calcification or replacement of the lamina by newly formed bone may take place. The contiguous ribs may be involved. Viterbo (1948) reported the occurrence of a large osteolytic bone defect in the right half of the body of the first lumbar vertebra, with no evidence of deformity or gibbus formation and normal intervertebral discs in a thirty-two-year-old woman with vertebral echinococcosis. Following immobilization, repair occurred, and re-examination showed in addition a paravertebral mass containing calcification and sclerosis. Schlanger and Schlanger (1948) mentioned that echinococcosis infestation of the spine is relatively frequent. Although the *x*-ray signs are insignificant at the beginning of the disease, clinically it may produce alarming symptoms originated by extension and infiltration of the meninges by the cysts. Spinal blocks may occur early in the disease and may be demonstrated by myelographic examination. They mentioned that the initial stage may present a partially destructive area in a transverse process far below the area of myelographic block, together with small clear bubble-like areas of radiolucency of the adjacent vertebral bodies. If the disease continues, destruction increases and the bubble-like character of the bone lesion disappears, being replaced by a destructive process. Spontaneous or operative fistulas may appear and obscure the bone picture. A widening of the interpedicular spaces in a case of hydatid disease of the twelfth thoracic vertebra was described by Oosthuizen and Fainsinger (1949). In their case myelography also disclosed a local block. They noted a hollowing out of the posterior surface of the involved vertebral body and observed that the findings could not be distinguished from those produced by any other expanding lesion in a similar situation.

In 4 of the 5 cases reported by Woodland (1949) there were symptoms of spinal cord pressure. The original site of the hydatid infection was the upper lumbar spine in 3 and the midthoracic spine in 2. No pathognomonic radiographic signs could be observed, but Woodland suggested that the absence of periosteal reaction minimal vertebral collapse and intervertebral disc narrowing slight sclerotic reaction with areas of destruction and involvement of contiguous ribs might be of some diagnostic aid. Rocca, Frano and Alayza (1950) reported a case of primary echinococcosis of the body of the sixth thoracic vertebra with cord compression and recovery after operation.

The differential diagnosis of hydatid disease of the spine, particularly in the presence of central nervous system derangements, is quite difficult. Included in consideration must be tuberculosis, malignant disease, neurofibroma, hemangioma and large perineurial sacral cysts. Myelography is important in determining the extent of encroachment into the spinal canal, and surgical relief may become necessary.

Schistosomiasis (*Bilharziasis*).—This condition is caused by any of three blood flukes. These may produce vesical or urinary symptoms (Schistosoma hematobium), intestinal manifestations (Mansoni), or may be of the oriental intestinal type (japonicum). The disease is due to contact with water contaminated with cercariæ discharged by infested snails. The primary infection occurs through the skin with distribution by way of the blood. Infection of the vertebral column by this disease is rare, but a case was mentioned by Gama and Marques de Sa (1945) in a forty-two-year-old male who had pain in the back and paralpegia. A myelographic examination showed partial block at the level of the sixth dorsal vertebra and complete block at the level of the twelfth dorsal vertebra. At operation an intramedullary granuloma containing eggs of Schistosoma mansoni was found.

Cysticercus.—This follows ingestion of raw or partially cooked pork infected with Cysticercus cellulosa. Infection may also occur through ingestion of contaminated water or food, particularly uncooked vegetables. The clinical aspects and diagnosis of spinal cysticercus was described by Bärtschi-Rochaix and de la Cuadra (1946), who reported that the symptoms in their cases simulated intracranial or spinal disorders. In 1 case, a laminectomy from the third to the fifth thoracic segments showed spinal cysticercus in the excised tissue. A primary meningeal intracranial reaction was also associated in this case with a severe and widespread spinal arachnoiditis. The diagnosis of spinal arachnoiditis was made by the characteristic findings on spinal tap and myelography. The etiologic diagnosis was confirmed by microscopic examination of the material removed at operation.

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"IVORY VERTEBRÆ"

This condition is one in which a distinctive radiologic appearance may be associated with various etiologic factors, all quite different. The usual roentgenographic picture is a vertebra with a dense, white appearance in contrast to the normal or possibly osteoporotic appearance of the remainder of the vertebral column. Such changes have been associated with Paget's disease, with myelomas and osteosclerotic metastases, particularly from carcinomas of the prostate, and with Hodgkin's disease. The association of so-called ivory vertebræ with post-traumatic changes in which there is first an osteoporosis followed by a sclerotic process was mentioned by Schinz (1952) and his co-workers as a progressive change in Kümmel's disease. Bársony and Schulhof (1930) believe the sclerotic changes might be a reaction to a low grade infectious process. We have recently had a 16-year-old male patient who had a classically dense, ivory vertebræ caused by a staphylococcic infection verified by culture of bone obtained at biopsy.

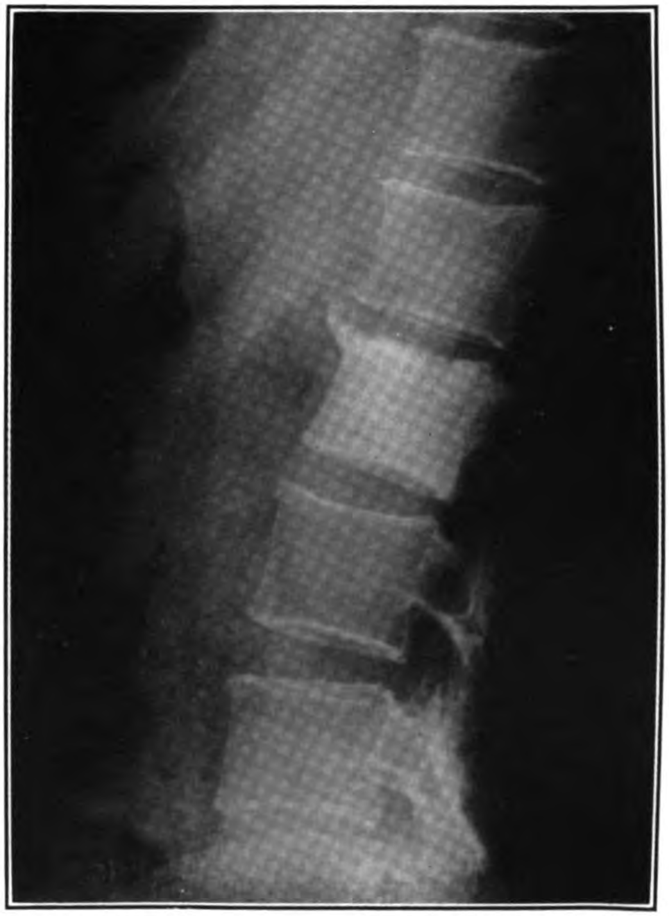

FIG. 174.—Ivory vertebra, later proved to be due to Paget's disease.

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CALCIFIED MEDULLARY DEFECTS

These were described by Ferguson (1947) as solitary rounded areas made up of irregular nodules of amorphous calcification, in or near the metaphyseal regions of the long bones, frequently in the center of the medullary cavity. These measure from 1 to 2 cm. in diameter, and as a rule, are solitary lesions which are sharply demarcated from the adjacent reticular bone. Fergusion believed that the lesion represented degenerated islands of cartilage which had become calcified. The solitary non-symmetrical nature supported the idea that trauma may be an etiologic factor. Steel (1950) also described these islands of bone, mentioning that they were homogenous in density, varying from 0.5 to 7.5 cm. in diameter. In a review of 6,000 roentgenograms, 120 calcified islands were found, most often in the femoral condyles, the iliac wings, the proximal portions of the tibia and head of the humerus. According to him, the etiology was not established, but 27 per cent of the lesions were found in patients suffering from blood vessel abnormalities. The possibility of bone infarction was strongly considered by him.

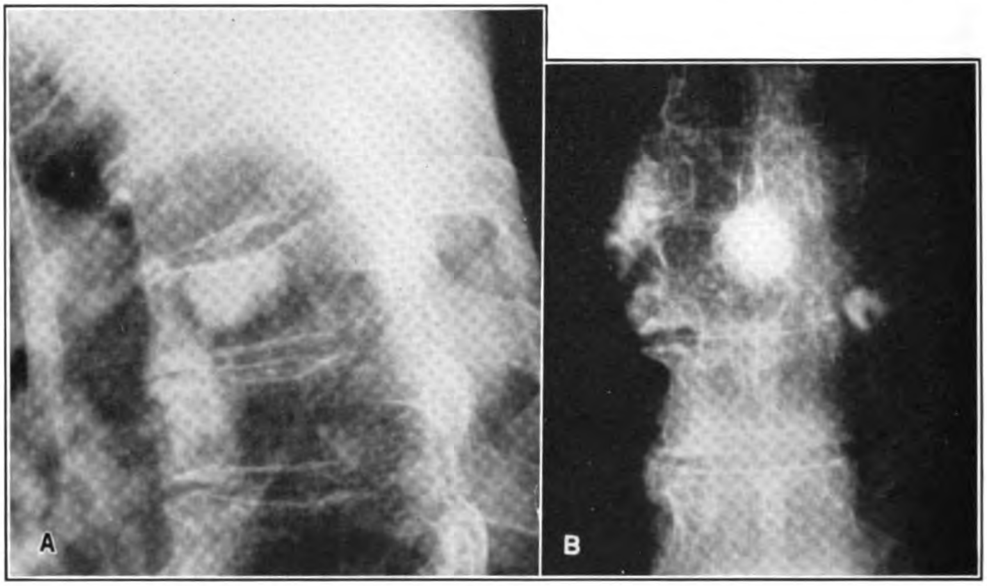

FIG. 175.—Localized enostosis in the body of the seventh thoracic vertebra. Incidental finding made during course of chest examination.

In our own material we have encountered 7 cases of sclerotic bone islands within the vertebral bodies. Two of these are illustrated here (Figs. 175 and 176). These varied in size and were irregular, slightly lobulated dense sclerotic foci of roughly oval shape, measuring up to 3.5 cm. in their widest diameter. As a rule, they were sharply delimited from the adjacent cancellous bone, and did not distort the outlines of the vertebral body. The lesions were solitary and were encountered as incidental findings. They were not associated with any specific symptoms. We have had no opportunity to confirm their presence at necropsy, so that histologic details are lacking. However Ackermann and Schwarz (1958) were able to demonstrate the benign sclerotic nature of this condition on biopsy material.

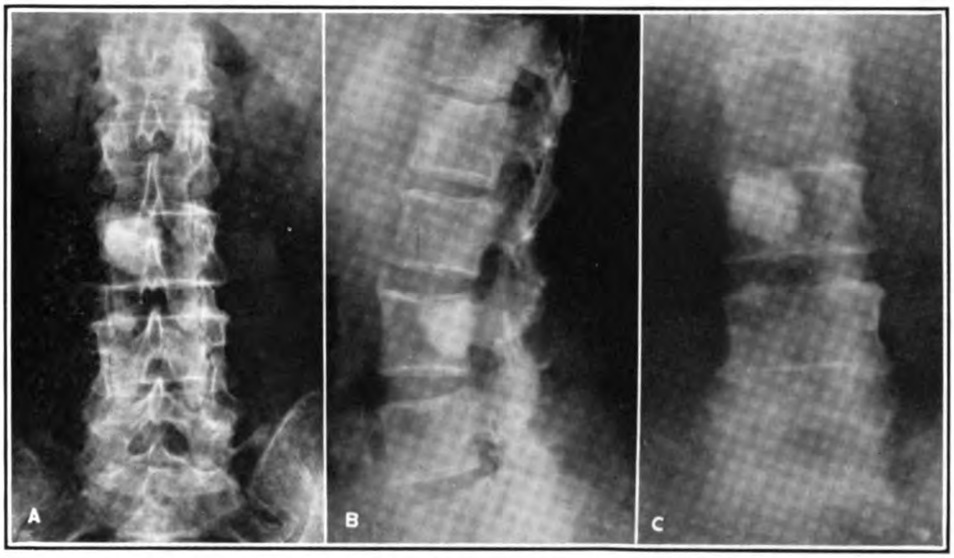

FIG. 176.—Calcified medullary defect in body of the third lumbar vertebra. Anteroposterior, lateral and anteroposterior body section roentgenograms.

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CAISSON DISEASE

Caisson disease is due to an accumulation of nitrogen gas in the blood vessels within the bones, with resultant aseptic necrosis. The common sites for involvement are the head of the femur or humerus. The necrotic lesions may break down and be invaded by, or eventually replaced by, new bone or fibrous tissue. Medullary calcifications in the diaphyseal ends of the long bones and secondary hypertrophic arthritis may appear as the disease proceeds to healing (Phemister, 1940). Rendich and Harrington (1940) encountered no spinal involvement in 4 cases. Coley and Moore (1940) suggested that the reason the vertebræ escape in this condition is the high proportion of red as compared to fatty marrow. They pointed out that the tissues with the poorest blood supply in relationship to nitrogen content are the bone marrow and the spinal cord. Those tissues with high nitrogen content are enclosed in bone, thus preventing free diffusion of nitrogen into contiguous tissues. This makes them dependent upon a comparatively poor blood supply for absorption and elimination. Consequently nitrogen elimination from these tissues is retarded when bubbles are present in the blood.

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OSTEITIS CONDENSANS ILII

Although not a specific disease of the vertebral column, its close association with the sacro-iliac region makes inclusion desirable at this point. This benign, self-limited condition occurs mostly in females of the childbearing age, and may be associated with recurrent attacks of low-back pain. Roentgenographic examination reveals a condensation of bone in one or both iliac bones adjacent to the sacro-iliac joints. Gillespie and Lloyd-Roberts (1953) encountered this condition in 2.2 per cent of 760 lumbosacral spines, and noted sacral as well as iliac bone involvement. In 1 case with histologic verification they found that the bone was harder than normal and bled less readily. The bone tissue showed no qualitative abnormality, but was gradually thickened by deposition of lamellar bone of normal structure. The ligaments and cartilage were normal. These writers postulate a vascular deficiency as a cause, with resultant sclerosis. Rendich and Shapiro (1936) reported 1 case in which a fusion of the sacro-iliac joint was done and a biopsy of the involved bone was obtained. This presented condensation of osseous tissue with obliteration of the evident former lacunæ. There appeared to be no osteolytic or osteoplastic change in the bone.

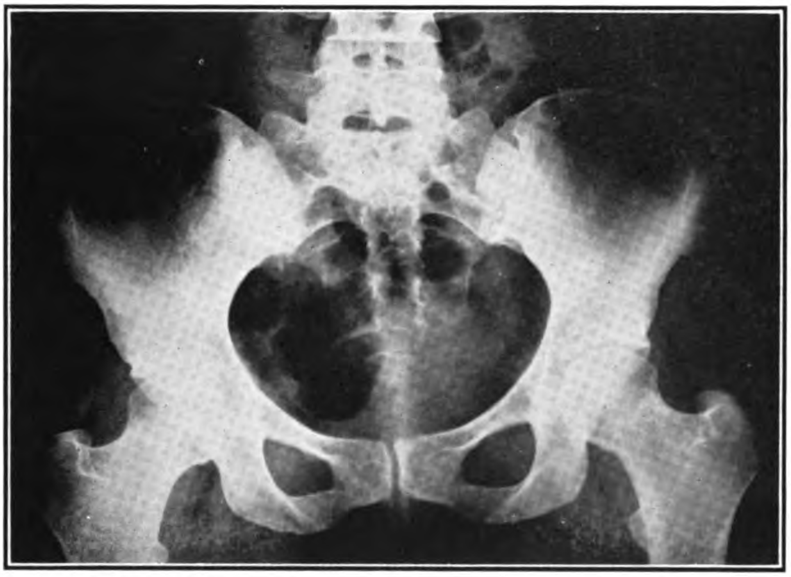

FIG. 177.—Osteitis condensans ilii involving right sacroiliac region.

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PAGET'S DISEASE (Osteitis Deformans)

This disease was originally named osteitis deformans by Sir James Paget in 1806, because he believed that it represented a mild, progressive inflammatory lesion of bone of an unusual nature. This etiology is no longer considered acceptable, and it is generally conceded that the origin of Paget's disease is unknown. The histologic appearance of the affected bone was well described by Schmorl (1931, 1932), who stressed the so-called mosaic structures consisting of many irregular islands of newly-formed bone cemented together by heavily staining lines, indicating rapid absorption and still more extensive and hasty reformation of bone. These rapid sequences of bone resorption and regeneration eventually lead to a secondary medullary fibrosis. The pathologic appearance of involved vertebræ is quite variable. The cancellous tissue becomes thickened and whitish, compressing the marrow cavity. Progression of the disease is accompanied by softening of the bone and progressive deformity. The initial phases occur in small isolated foci consisting of thickened trabeculæ. Productive changes follow the initial softening and the end result may, in isolated cases, be a so-called ivory vertebra in which the entire body assumes a dense, white appearance as seen radiologically. This appearance is not in accord with the actual density of the bone, which may be relatively soft and compressible, and entirely unlike ivory in its consistency.

Paget's disease may occur in a monosteitic form or as a diffuse disease. Its occurrence in a single vertebra was commented on by Schmorl, who accepted it as a variety of the condition. Bársony and Schulhof (1930) produced evidence to show that so-called ivory vertebræ were local manifestations of Paget's disease. Much more common, however, is the appearance of Paget's disease in many vertebræ. Usually the lumbar spine and sacrum are involved to the greatest extent, the dorsal spine next in frequency, and the cervical spine is rarely involved.

Compression of the spinal cord in Paget's disease was reported by Wyllie (1923) in 4 cases. Colclough (1949) was able to collect a total of 27 cases up to 1948, and added 2 of his own. Nineteen had been operated upon with varying degrees of return of function. The point of exact block can be discovered by myelography. A case of Paget's disease in which compression of the spinal cord was caused by hypertrophied laminal arches of the first, second, and third dorsal vertebræ was reported by Teng, Gross, and Newman (1951). An unusual case of Paget's disease affecting the first and second cervical vertebræ with spinal cord compression was described by Whalley (1946) in a sixty-six-year-old man in whom there was a spontaneous forward dislocation of the head on the cervical spine. Roentgenograms showed massive new bone formation, fusion of the vertebræ, kyphosis, and dislocation of the head, with narrowing of the spinal canal. At necropsy there was encroachment of the bony overgrowth on the spinal canal and its contents. Dickson, Camp and Ghormley (1945) mentioned that in their series there was evidence of compression of the cord following pathologic fracture of the vertebræ in 6 cases. Laminectomy had been performed 4 times on 1 patient in a period of five years. Myelographic block was encountered in 3 cases reported by Latimer, Webster and Gurdjian (1953).

The presence of malignant degeneration of Paget's disease is well known. Its occurrence is mainly in the long bones and pelvis, occasionally in the skull, and infrequently in the spine. Campbell and Whitfield (1943) reported that in a search of the literature they encountered but 3 cases of sarcoma of the vertebræ, in all of which involvement of other bones was present so that it was difficult to determine the primary lesion. Their search revealed an incidence of osteogenic sarcoma of various bones in Paget's disease of from 7.5 to 14 per cent according to different authors. In 3 cases reported by them compression of the spinal cord and cauda equina was associated with severe pain and neurologic changes. Summey and Pressly (1946) collected 76 cases of sarcomatous degeneration of Paget's disease of bone from the literature, of which 9 were in the vertebral column, almost always in conjunction with changes elsewhere in the skeleton. Paget's sarcoma is highly malignant, may be multicentric in origin and is frequent in situations where the original disease is infrequent, as in the humerus. In bones commonly affected, such as the pelvis or vertebræ, sarcomatous degeneration is relatively uncommon.

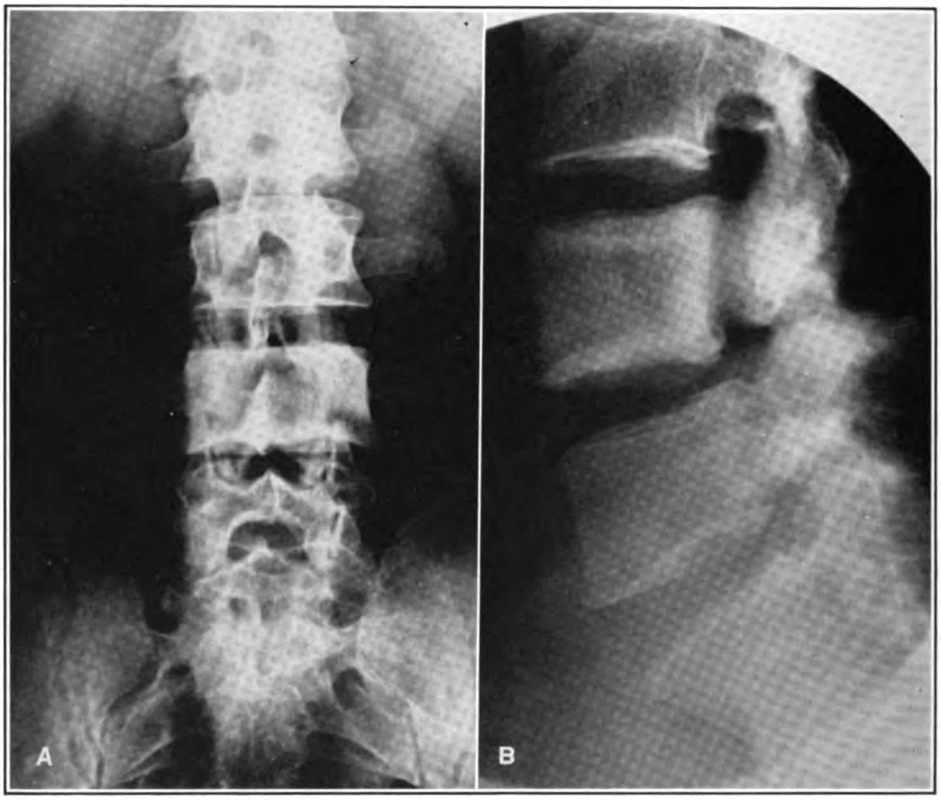

FIG. 178.—*A* and *B*, Early changes of Paget's disease in body of fourth lumbar vertebra. The pedicle and neural arch also are thickened and increased in density. Note the coarsening of the cancellous bone in the vertebral body, surrounded by a thickened periphery.

The radiologic diagnosis of Paget's disease is often made incident to other roentgenographic examination in which the vertebral column happens to be visualized. In many the disease is entirely unsuspected. The earlier stages may be manifested by a slight diffuse increase in the density of an involved vertebra which in some instances presents vertically parallel thickened trabecular markings. These may occupy the entire vertebral body or in some instances are limited to a relatively small portion of the vertebra. The changes are most frequently seen in the vertebral body but sometimes occur in the neural arch as well. Involvement of the spinous and transverse processes in early cases is relatively uncommon. In other instances the disease manifests itself by the presence of small, somewhat circular shadows of increased density within the vertebral body. As a rule, these are multiple and in early stages are unaccompanied by any changes in the configuration of the vertebræ. As the disease advances there may be a mild flattening of the involved vertebra, sometimes with bulging of its borders. A diffuse increase in the density of the vertebral body approximating the cotton wool appearance usually associated with Paget's disease in the flat bones may appear. Mention has already been made of the monosteitic form of Paget's disease involving a single vertebra. Our experience has been that this is relatively uncommon. Not infrequently both the thoracic and the lumbar spine are involved at the same time. In the more advanced forms of the disease the vertebræ may assume an almost flattened elongated appearance. In these instances involvement of the neural arches

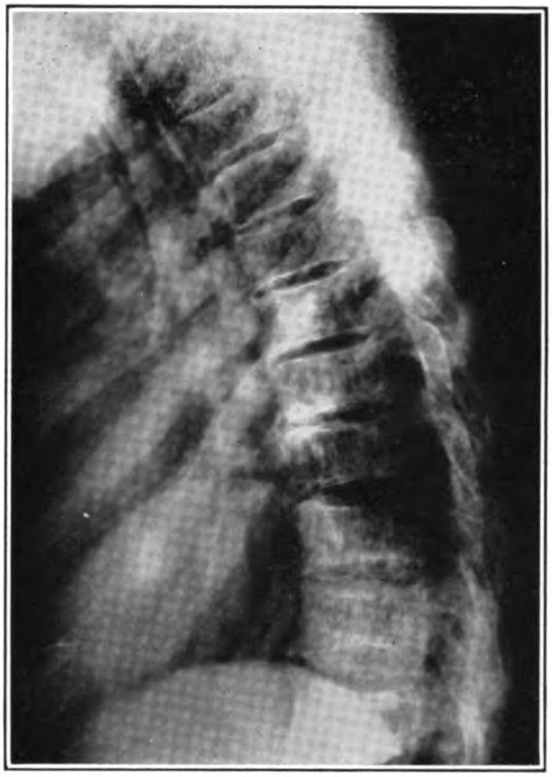

FIG. 179.—Scattered osteitis deformans involving multiple thoracic vertebræ.

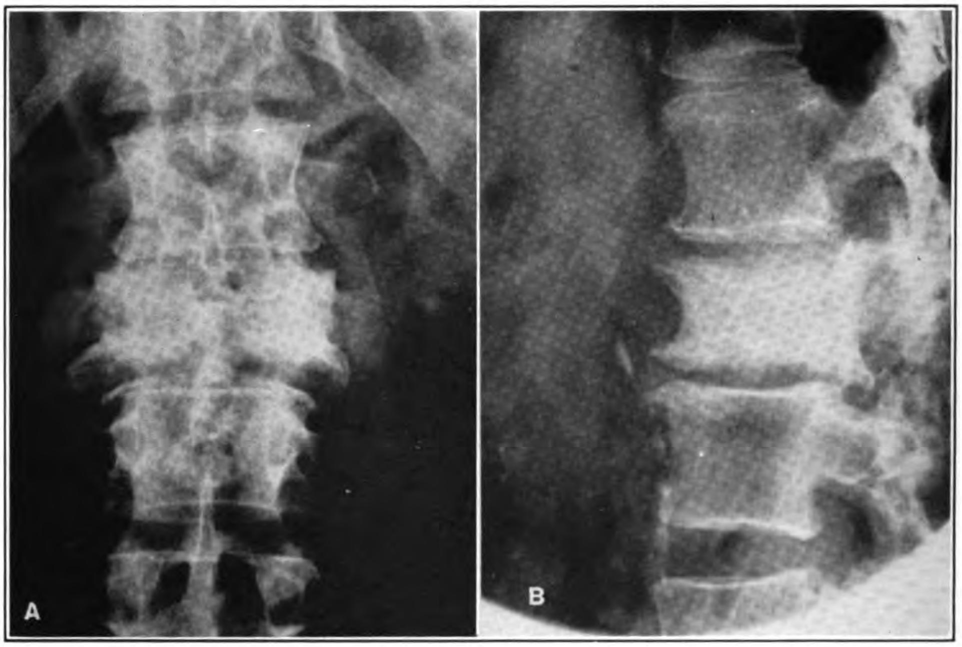

FIG. 180.—Paget's disease of second lumbar vertebra, with compression and expansion of the vertebral body, and spur formation laterally and posteriorly.

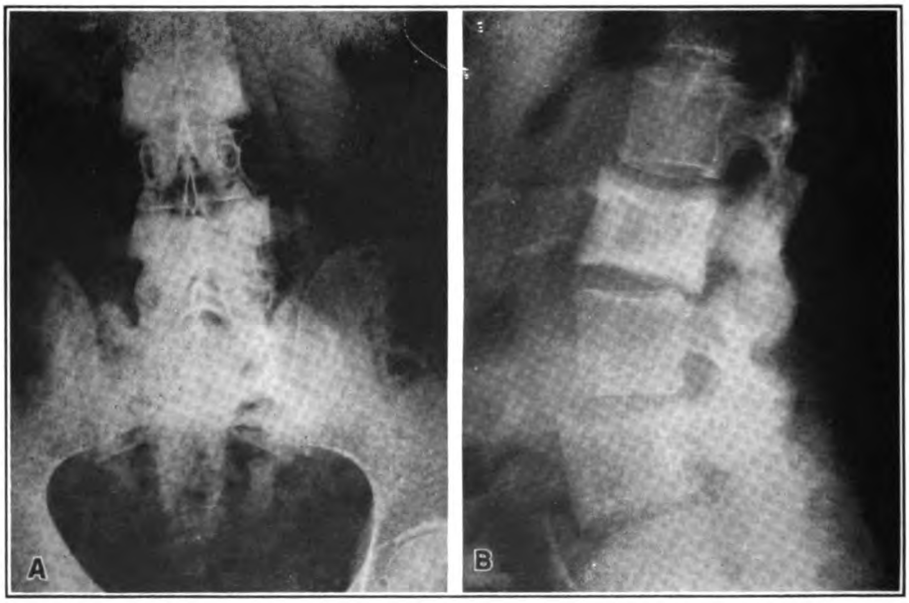

FIG. 181.—Paget's disease of the lumbar vertebræ, with expansion and increase in height of the second and fourth lumbar vertebræ.

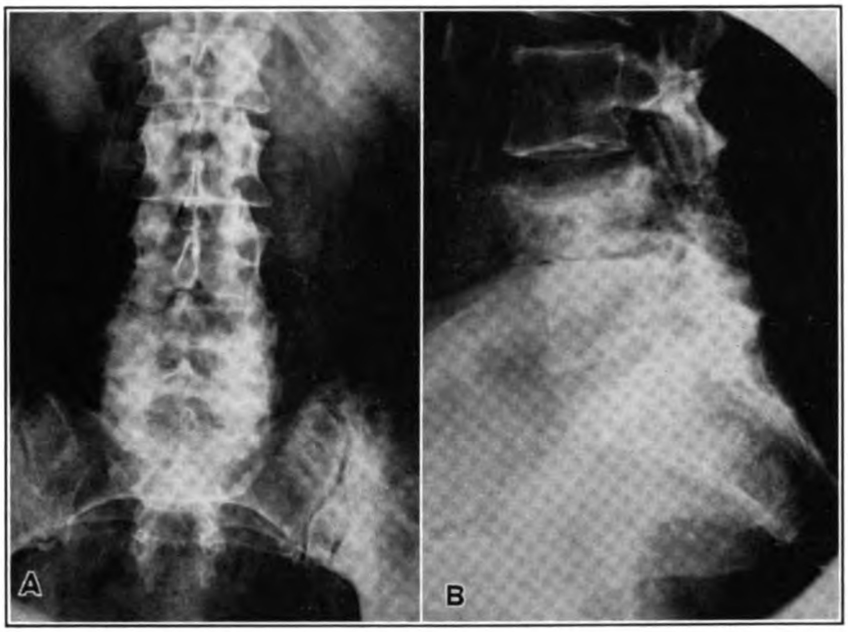

FIG. 182.—Paget's disease of the fourth lumbar vertebra, with compression and predominantly lytic changes.

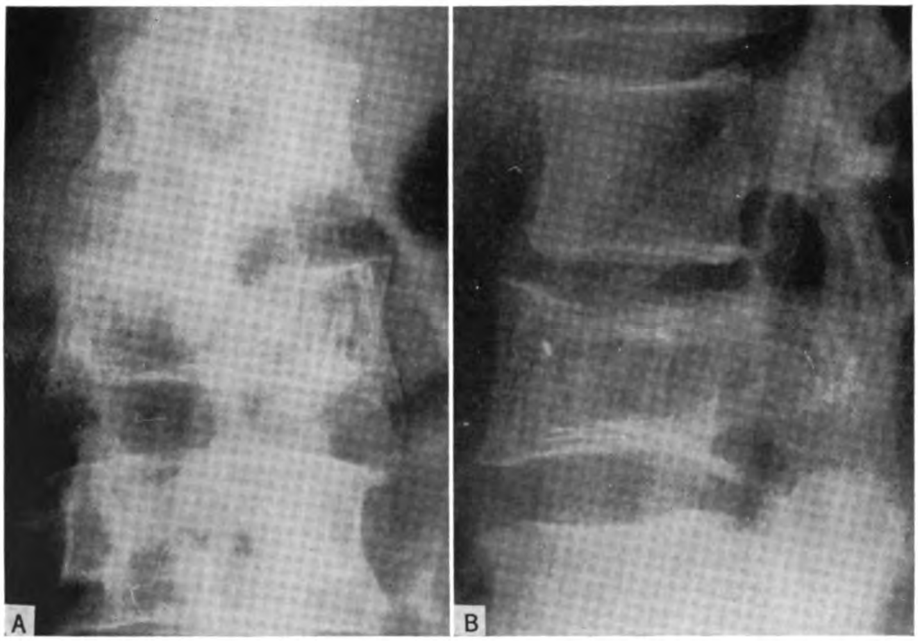

FIG. 183.—Monostotic Paget's disease involving second lumbar vertebra, lytic variety. Note the spreading of the vertebra and the indentations into its upper and lower surfaces. The pedicles laminæ and transverse processes are involved.

may be relatively common. Deformity of the intervertebral discs is no more common in patients with Paget's disease than is observed in the similar age groups. However, infrequently one may observe a loss of density of a vertebral body with encroachment of the intervertebral fibrocartilage into the involved vertebra with a consequent loss of the height of that vertebral body. In 1 of our cases the intervertebral space between the fourth and fifth lumbar vertebræ practically disappeared into the degenerated softened inferior aspect of the fourth lumbar vertebra.

Snapper (1951) mentioned that in patients over forty years of age, with hypertrophic changes in the spine, Paget's disease starts in the osteophytic proliferations and then gradually spreads to the vertebral bodies.

One may sometimes encounter difficulty in evaluating the status of a patient suspected of Paget's disease purely from the spinal roentgenograms. In such cases investigation of the other bones, and particularly of the pelvis and skull, may prove of great importance.

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RADIATION EFFECTS

The effect of intensive *x*-ray irradiation on the vertebral column has been known for a long time (Desjardins, 1930; Warren, 1943). Smithers, Clarkson and Strong (1943) reported radiation myelitis in a patient who received a depth dose of 5800 r in thirty-nine days for carcinoma of the esophagus. The lesion developed fifteen months later, presenting a Brown-Séquard syndrome which they considered to be due to intramedullary gliosis. Radiation myelitis of the brain stem was reported by Boden (1948) as occurring in 5 fatal and 5 nonfatal cases in a series of 161 patients who had received *x*-ray therapy for malignant tumors of the nose, mouth, larynx or cervical lymph nodes, the myelitis appearing in the cervical spinal cord. A long time interval occurred between the appearance of symptoms and the cessation of treatment. Initial symptoms were numbness and tingling of the hands and feet, precipitated by flexion or extension of the neck, and often associated with shoulder or neck pain. In a few cases the symptoms were transient, disappearing in from three to nine months. In the others parasthesias were followed by weakness or spastic paralysis of one or more extremities, and later bowel or bladder dysfunction. In the fatal cases there was intercostal paralysis and death from bronchopneumonia or urinary tract infection. Greenfield and Stark (1948) also reported on post-irradiation neuropathy in 3 cases where neurologic disability followed intensive million-volt *x*-ray therapy to the retroperitoneal region. In each case a flaccid paralysis appeared, and the chief site of injury was believed to be the anterior horn cells in the lumbosacral segments of the cord. No histologic material was available. These authors calculated the doses to be from 5000 to 6000 r, delivered to the retroperitoneal areas for nodes from primary testicular tumors. The *x*-ray examination and myelograms of these patients were all within normal limits. Seven cases of fatal damage to the brainstem after *x*-ray therapy for malignant tumors of the middle ear, pharynx or parotid gland were reported by Boden (1950) who concluded that the limited tolerance of the central nervous system was from 3500 to 4500 r, depending on the size of the field used. The *x*-rays affected primarily the blood vessels rather than the neurones, and the eventual neurologic lesions resulted from impaired circulation. Radiologic examination in these people showed no evidence of bone change.

Radiographic evidence of destruction of bone following *x*-ray therapy was reported by Spitz and Higginbotham (1951) in a forty-six-year-old man who developed an osteogenic sarcoma following irradiation for testicular seminoma. Roentgenograms of this patient showed destruction of the laminæ of the third, fourth and fifth lumbar vertebræ with calcific flecks in the soft tissues, and distinct bulging of the right psoas shadow. Extraskeletal osteogenic sarcoma arising in irradiated tissue was also reported by Auerbach, Friedman, Weiss and Amory (1951) in a case of testicular tumor following intensive treatment. Necropsy revealed postradiation effects and a huge osteogenic sarcoma of the right lumbar muscles, with invasion of adjacent vertebræ and ribs, and lung and lymphatic metastases. Four years had intervened between *x*-ray therapy and the clinical onset of the malignant disease. The roentgenograms showed a calcific mass in the soft tissues, not connected with bone or skin.

Retardation of bone growth, with deformity of muscular bony architecture, atrophy, and scarring of the skin and fibrosis of subcutaneous tissues were reported by Murphy and Berens (1952) as a late sequela following heavy irradiation in children. These authors commented that changes usually cannot be avoided if a clinical cure is to be attained. In a detailed report on the irradiation effects of roentgen therapy on the growing spine Neuhauser, Wittenborg, Berman and Cohen (1952) described 45 patients who had had *x*-ray therapy over the spine, together with postmortem examinations of 11 of these. These authors suggested, as a preliminary guide, that dosages under 1000 tissue roentgens usually fails to produce a gross permanent defect in a growing child, irrespective of the age at which treatment is given. Children over two years of age may tolerate between 1000 and 2000 r delivered to the spine, with only minor growth disturbances, but dosages in excess of 2000 r, irrespective of age, will probably produce growth disturbances. In their series these were associated with gross abnormality in the contour of the vertebræ. Scoliosis was rarely produced if the spine was uniformly irradiated. The significant changes in the eleven post-mortem examinations were nonspecific growth retardation and irregularity of ossification of the epiphyseal cartilage. These changes may lead to an irregular advance of the line of ossification, but no alterations of osseous contour were described. Benign cartilaginous exostoses of the types seen in multiple and familial form appear to arise with greater frequency in bones where epiphyses had been irradiated than in the normal population. Their findings were much the same as those described by Arkin and Simon (1950) in their experimental study of the production of scoliosis by irradiation in which vertebral epiphyseal growth was suppressed by irradiating the vertebral bodies asymmetrically. Whitehouse and Lampi (1953) followed 4 patients who had been treated for renal tumors in infancy and childhood for ten years, and described vertebral damage which they associated with dosage factors.

The effects of ingestion of radium were described by Stevens (1942) in a thirty-six-year-old man who in 1926 was described as having Hodgkin's disease and as a heroic measure had been given seven injections of radium chloride intravenously. The first injection was 20 micrograms, followed by 6 injections of 50 micrograms in 1926, 4 injections in 1927 and 1 each in 1929 and 1930. In this case radiation necrosis of the jaw, with sequestration of bone containing radium, followed. Destructive changes appeared in the dorsal vertebræ before the mandibular destruction became apparent.

The histology of irradiation sarcoma does not differ from spontaneous osteogenic sarcoma. The tumors are essentially of the pleomorphic spindle cell variety. In many giant cells are prominent, and cartilaginous and myxomatous changes are not uncommon (Jones, 1953).

It has long been known that even though the brain and spinal cord were relatively resistant to the effects of irradiation, dosage of sufficient quantity could result in damage. Radiation myelitis has been reported in patients who have been subjected to supervoltage treatment for various malignant lesions in which the spinal cord had been included in the field. It has been estimated that doses beyond the 5,000 r range are more likely to be followed by a radiation reaction. The damage produced by the roentgen rays affects mainly the vascular structures in the spinal cord, resulting in occlusion because of vascular thickening with enlargement and swelling of the endothelial cells followed by a fibroblastic reaction. The symptoms of radiation myelitis appear slowly, the time interval varying from about seven months to well over three years. According to Dynes and Smedal the average interval between completion of treatment and the development of myelitis is well over 23 months. The symptoms are those of any other type of myelitis, with paresthesias, sensory levels and evidences of long tract involvement of both the motor and sensory pathways. In some patients the symptoms may be transitory and then vanish, but where the damage has been more extensive the final result, depending on the location and extent of the changes, may be paraplegia or quadriplegia.

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FLUORIDE POISONING

This condition has been described in individuals who have been in contact with various forms of fluorides, such as cryolite miners as described by Møller and Gudjonsson (1932) who reported widespread osteophitic formations at the attachments of ligaments, tendons and periosteum together with widespread osteosclerosis. Cryolite is the double fluoride of sodium and aluminum, containing as much as 54 per cent of fluorine. Bone changes in chronic fluorine intoxication were reported by Bishop (1936) in a laborer who for 18 years had been exposed to the dust of fertilizer made by crushing a phosphate rock which had a high fluorine content. Sclerosis of the entire skeleton except the skull was noted, with marked increase in density of the bones, together with vertebral osteophytes and calcifications of the attachments of some of the ligaments. In a chemical analysis of the bone in Bishop's case, Wolff and Kerr (1938) found an increase in fluorine content up to 20 times that of normal. The fluorine content was highest in the vertebræ and in the long bones, and had been deposited as calcium or magnesium fluoride. Wilkie (1940) reported 2 cases of fluorine osteosclerosis which occurred secondary to exposure to hydrofluoric acid fumes over a long period of time.

A much more common form of exposure is that secondary to the consumption of drinking water of high fluorine or fluoride content, such as may occur in North Africa, India or Argentina. The literature on this was cited by Greenwood (1940), who reported fluoride osteosclerosis in such patients. The effect of drinking contaminated water was graphically described by Kilborn, Outerbridge and Lei (1950). This happened in a Chinese village in which the inhabitants drank water contaminated by fluorine, and consequently developed fluorosis. Pigs, which shared the human water supply, were also affected, but other animals which used pond water containing no fluorine escaped. These authors described the skeleton of an affected male who died at the age of thirty-seven and whose skeleton was available for study one year later. All the bones were markedly thickened, porous, rough and brittle. There was complete ossification of the intervertebral discs with fusion of the vertebræ making the spine one long rigid bone. Chemical study showed about 20 times the normal fluorine content for many of the bones with an even higher ratio for the teeth. Roentgenograms showed increase density, thickened, trabeculæ and narrowing of the marrow cavities.

In a report on the roentgenographic changes and urinary fluoride excretion among workmen engaged in the manufacture of inorganic fluorides Largent, Bovart and Heyroth (1951) mentioned that 22 cases had been reported in the American literature, including 6 in their paper. There appeared to be a correlation between the level of fluoride excretion in the urine and the presence or absence of changes in the bone density. These authors concurred with other observers in that the increased density of bone is particularly prominent in the vertebral column, but they noted that in their cases little or no hypertrophic changes existed. This observation was in accord with that made by Linsman and McMurray (1943) who had also reported a case of fluoride osteosclerosis from drinking water in which necropsy showed no evidence of osteophyte formation.

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Lead Poisoning

The lead line associated with ingestion of lead in growing children is well known. However such changes do not as a rule appear in the vertebræ, and in a review of 10 cases of lead poisoning in our own series the roentgenograms of the spine were normal even when the epiphyseal lines of the long bones showed dense lead lines.

Chapter

5

Neoplasms of the Vertebral Column

NEOPLASTIC METASTATIC DISEASE

The most common of the tumorous disorders of the vertebral column are metastatic neoplasms. Involvement usually occurs by extension through the blood stream, either the systemic circulation or through the venous vertebral plexus. Direct spread of a tumor originating in close proximity to the spine also may be observed. Invasion of tumor into regional lymph nodes is believed to occur by embolization through the lymphatics to the affected glands. The question as to whether lymphatic drainage is a wholly acceptable route for metastases to the vertebral column is discussed by Willis (1952). According to him lymphatics have not been demonstrated in the bone marrow, and cancerous permeation of the lymphatics in the deep fascia, periosteum and soft tissues adjacent to affected bones is rarely present. As a result of his own work and a review of the literature, Willis concluded that the association of lymph node metastatic deposits and metastases to adjacent bones was purely coincidental, and that the hypothesis of lymphatic entry of neoplastic metastases into bone should be abandoned. The role of the spinal plexus of veins in the spread of neoplasms to the vertebral column is best exemplified by metastases from prostatic carcinoma. Batson's work on the passage of radiopaque material into this system from the prostatic plexus has been mentioned in the first chapter. Willis, however, commented that while this may be one route for the spread of metastatic emboli, there was no reason to believe that remote metastases did not follow the usual hematogenous route. He went on to say, moreover, that freedom of the lungs from visible metastases in cases with bone extension is not paradoxical at all, and quoted the reports of Schmidt (1903) and other workers that microscopic study of the lungs in such cases shows the paradox to be illusory.

Direct extension of malignant neoplasms into the vertebral column is rather infrequent. The periosteum and cartilage maintain an effectual barrier against the penetration of tumors into the spine. When this barrier is disrupted by infection, however, neoplastic invasion of bone through the destroyed periosteum may readily occur.

Metastatic invasion of the vertebral bodies mainly occurs by the hematogenous route. As with other bones, the red marrow is almost always affected first. The tumor cells encroach into the spaces surrounding the bony trabeculæ, and later extend towards and destroy the cortex. As a rule, the periosteum of the vertebral bodies confine the tumor so long as it remains intact. If, however, there is periosteal destruction by local pressure, the tumors may extend beyond their confines and bulge into the adjacent tissues in much the same way as a paravertebral inflammatory process. As a rule, such neoplastic extension is less prominent than observed either with tuberculosis or pyogenic infectious.

Metastases frequently are described as being either osteoplastic or osteoclastic, the former referring to tumors associated with increased bony density, while the latter connotes osseous destruction and demineralization. Both may coexist not only in the same patient, but also in the same vertebra. The demineralization of bone associated with osteoclastic metastases is due to the fact that the tumor crowds out the calcium-bearing stroma. In more advanced cases, with obliteration of the vascular supply because of the packing of carcinomatous cells, necrosis of bone may occur with replacement of the neoplasm by degenerated and devitalized bone and metastatic tissue. The occurrence of increased bone formation with osteoplastic metastases has not been entirely explained. Willis regarded this increase as a reaction of bone to diffusable products from the tumor cells. He pointed out that various tumors possessed different intrinsic properties. For example, the highly osteoplastic nature of carcinoma of the prostate may be associated with a substance which causes rapid proliferation of bone. The presence of high serum acid phosphatase content may indicate that the substance is of enzymatic character. The association of osteoplastic metastases with slow-growing tumors such as scirrhous carcinomas of the breast, or prostatic carcinomas, and the association of osteoclastic metastases with

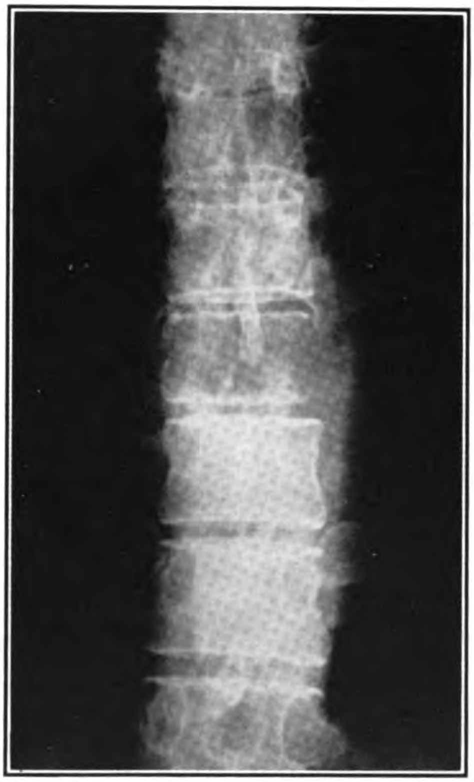
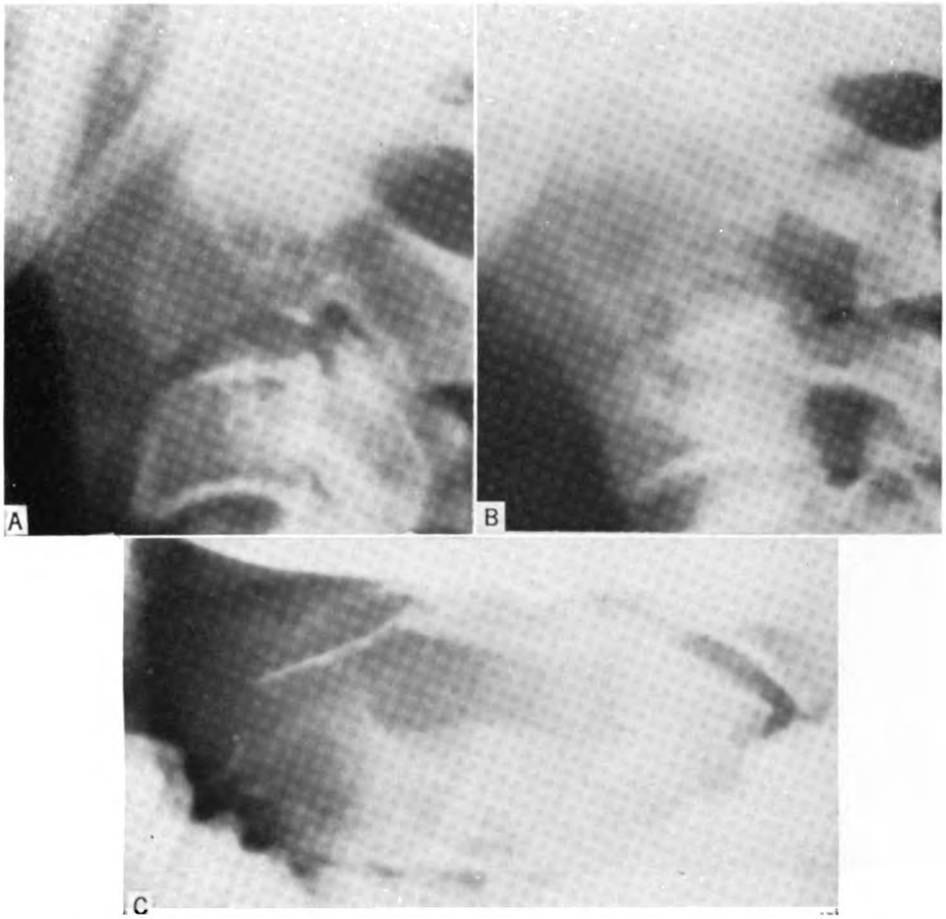


FIG. 184.—Metastases to the eighth thoracic vertebra from carcinoma of the breast. The pedicles are destroyed, the left completely and the right partially. The body of the vertebra is involved with a lytic process, but is not yet collapsed. Neoplastic tissue extends to the paravertebral soft tissue on the left side.

tumors of an anaplastic nature or with tumors deficient in stroma, such as thyroid or renal carcinomas, is of interest.

The usual symptom of spinal metastases is pain. However, it is well known that extensive skeletal metastases may exist without symptoms referable to the involved vertebræ. This occurs when the vertebral body maintains its configuration, and when there is no extension of the tumor to cause pressure on or invade the periosteum. Pain, to a large degree, is due to pressure on the richly innervated periosteum. In addition, pain may be produced by pressure on nerve roots or the spinal cord. The clinical picture sometimes suggests a herniated disc if a single nerve root in the lumbar spinal



FIG, 185.—Lytic metastasis to the first and second cervical vertebræ from an occult neoplasm of the lung. The patient had complained of pain in the back of his neck for about three months. No pulmonary symptoms were present, and the chest roentgenogram at this time was normal. The pulmonary neoplasm was diagnosed about four months after this examination. The body of the first cervical vertebra is extensively involved, and is destroyed more on the right than on the left side, as is the second cervical vertebra.

A, Film in extension shows the soft tissue swelling anterior to the first and second cervical vertebræ. *B*, In flexion the destruction of the second cervical vertebra is seen better. *C*, Open-mouth view shows the destruction on the right side of the first and second cervical vertebræ, and a shift towards the left side.

canal is impinged upon. Extension of a neoplasm higher in the spinal canal may be associated with a syndrome of compression of the spinal cord, the degree of which depends upon the pressure effect of the tumor. An interesting observation was made in several patients with severe pain in the vicinity of a vertebra in which marked destruction was evident radiologically, but in whom the vertebral body had retained its normal height. With collapse of the vertebra, and probably with lessening of tension on the periosteum, pain became less marked and in some instances actually disappeared.

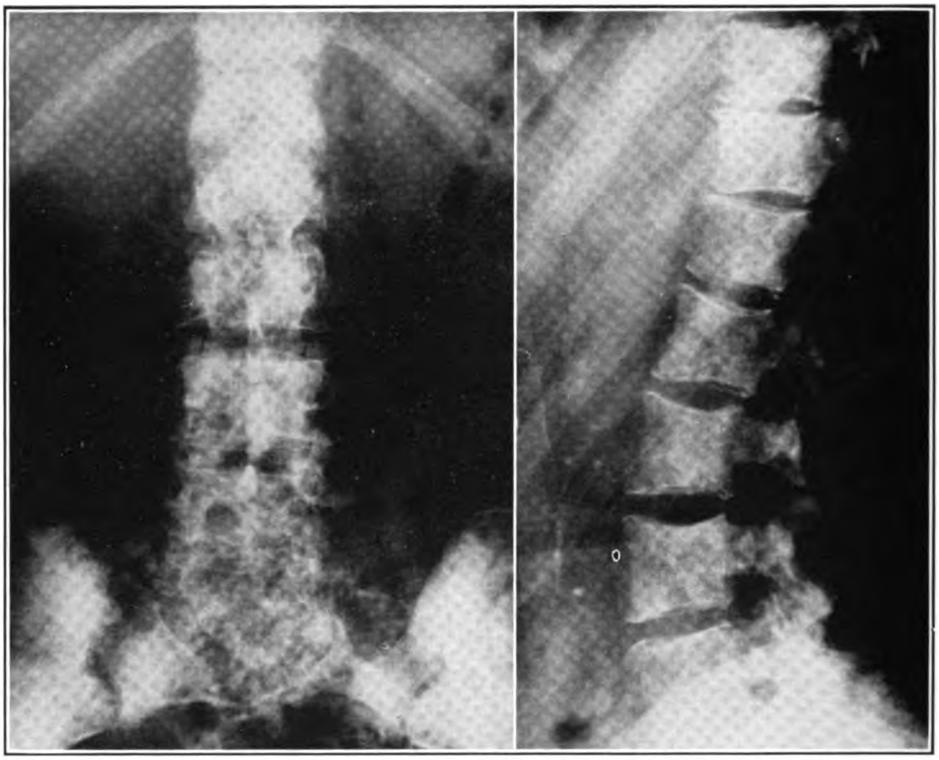


FIG. 186.—Anteroposterior and lateral roentgenograms of the lumbosacral spine showing mixed lytic and osteoblastic metastases from carcinoma of the breast.

The usual sites of origin of tumors which metastasize to the vertebral column are the breast, the prostate, the thyroid, the kidney and the lung. Less commonly tumors of the ovary, the uterus, the testes, the pancreas, biliary and the gastrointestinal tracts involve the spine. Melanomas not infrequently metastasize to the vertebræ. Other tumors are those of the bladder, the salivary glands, the lips, and various bone and soft tissue sarcomas. Of considerable importance is the occurrence of spinal metastases in patients with lymphoblastomas and leukemia. Another group of interest includes neoplasms of the brain which extend into the spinal canal by way of the cerebrospinal fluid circulation.

The radiologic identification of metastatic tumors in the vertebral column depends upon the identification of bone destruction or production. Not infrequently fairly large metastatic deposits exist with no visible disturbance in the contour or bony density of the affected vertebral segments. This was brought out by Snure and Maner (1937), who pointed out that cortical bone accounted for most of the detail of the bony structures depicted on roentgenograms, while the spongiosa accounted for the general bony density. Under these circumstances destructive changes in the marrow spaces, usually the first to be involved, easily go undetected until sufficient destruction of bone occurs to present itself on the roentgenograms as a shadow of decreased density. Their findings were borne out by Ardran (1951), who described experiments in which a 3-mm. hole drilled vertically through the center of a vertebral body could be seen only on a lateral roentgenogram and could not be identified on frontal films, where it was obscured by the spinous process. This hole was then filled with water and further roentgenograms under the same conditions as before showed that the loss of bone tissue could no longer be identified. Ardran then enlarged the hole by stages to approximately 14 mm. in diameter and roentgenologic studies showed that when these were filled with water the defect was only slightly visible as a porosed area. He concluded that an area of destruction of bone over 1 cc. in diameter in the center of a vertebral body may exist without definite *x*-ray evidence of its presence, and he noted that this comprised approximately 30 per cent of the bone thickness at this site. Further experiments were performed in which slices of bone were removed from the

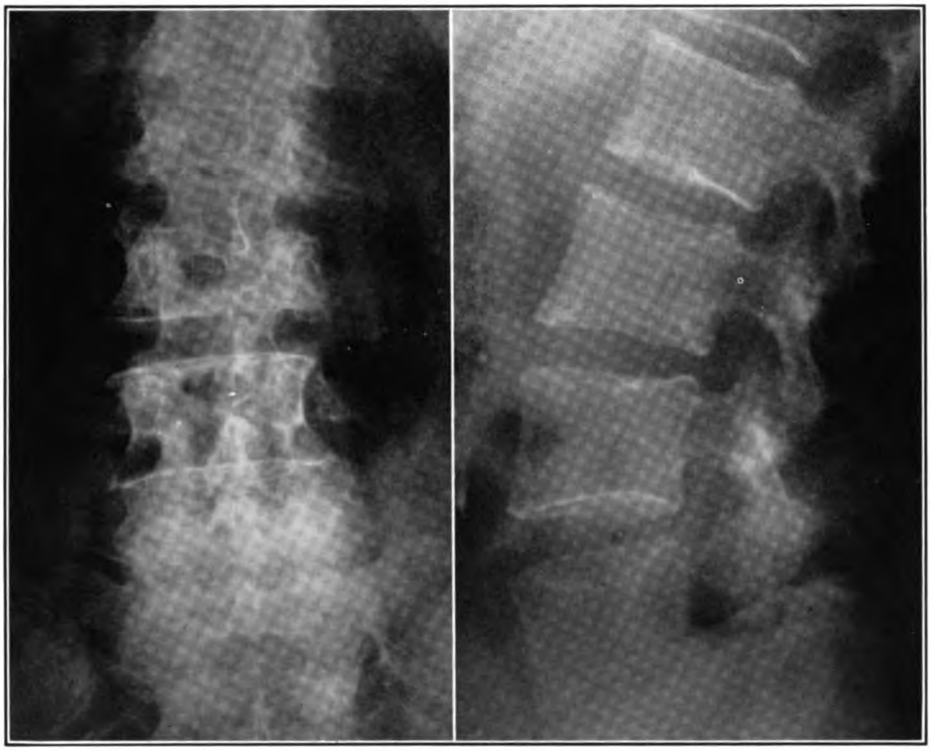


FIG. 187.—*A*, Metastases from carcinoma of the breast. The left superior aspect of the body of the third lumbar vertebra is compressed. *B*, Lateral view. There is no compression of the third lumbar vertebra, but its superior and inferior aspects have lost their normal osseous densities, and faint lytic changes are seen in the cancellous bone.

anterior aspect of a vertebral body parallel to the anterior spinal ligament in which over 1 cc. of bone had to be removed before the deficiency became apparent on the anteroposterior view.

The earliest roentgenologic evidence of metastatic disease is slight somewhat spotty decalcification of a vertebral body, often associated with a slight change in its contour. By the time this occurs the metastasis usually is quite large and of sufficient extent to have disrupted the spongiosa sufficiently to permit some collapse of bone. Usually this is associated with some extension to the cortex from the inner aspect of the vertebral body, weakening the buttressing effect of the solid portion of the vertebral body. Nevertheless, from the roentgenographic viewpoint the thing to watch for is slight decrease of density associated with the minimal changes in the configuration of a vertebral body. This may be so slight as to involve only a centimeter or two and sometimes may be seen only on one projection, either frontal or lateral, depending upon the location of the destroyed bit of bone. Less frequently particularly where there is an osteosclerotic reaction such as may occur with carcinoma of the prostate, the initial indication of a metastic deposit may be a small somewhat circular, usually poorly defined area of increased density only a few millimeters in diameter. In occasional cases, particularly in metastases from breast tumors, areas of radiolucency may coexist with small spotty areas of increased bony density.

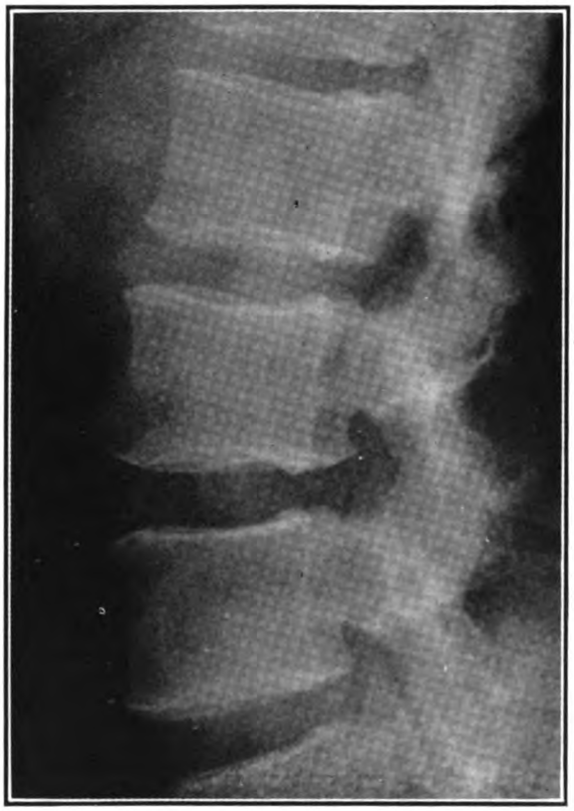


FIG. 188.—Metastatic destruction of the anteroinferior aspect of the body of the third lumbar vertebra seen on the lateral view only.

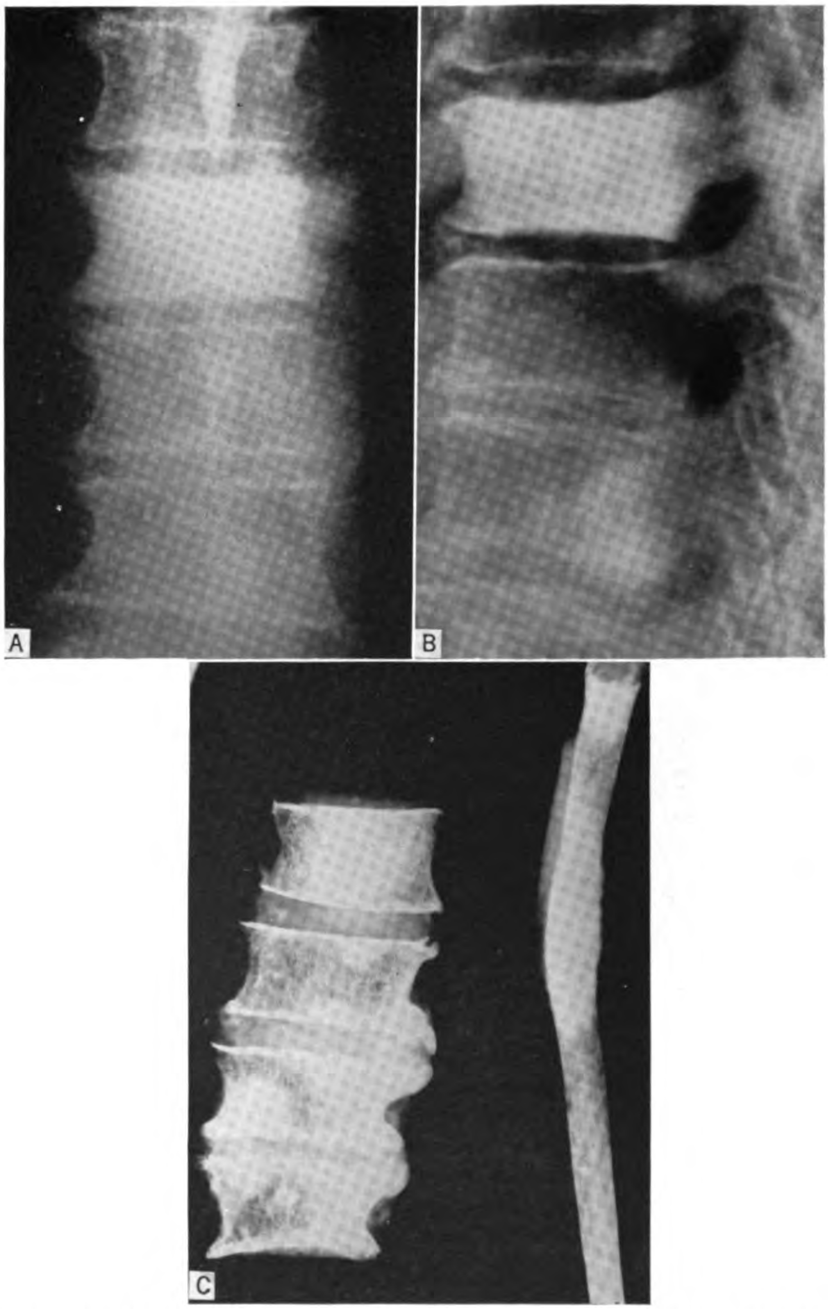


FIG. 189.—*A* and *B*, Anteroposterior and lateral roentgenograms of midthoracic vertebræ with densely sclerotic metastases from a prostatic carcinoma. *C*, Roentgenogram of vertebral slab and one of the adjacent ribs.

With progression of metastatic involvement and increasing weakness of the vertebral body, distortion follows either as a diffuse flattening of a vertebra or not infrequently, the destroyed region may involve either half or the anterior aspect of a vertebral segment. It is not uncommon to observe a vertebra in which one lateral half is diminished considerably in height, and the zone of distinction between the destroyed area and the uninvolved bone may be fairly sharply demarcated.

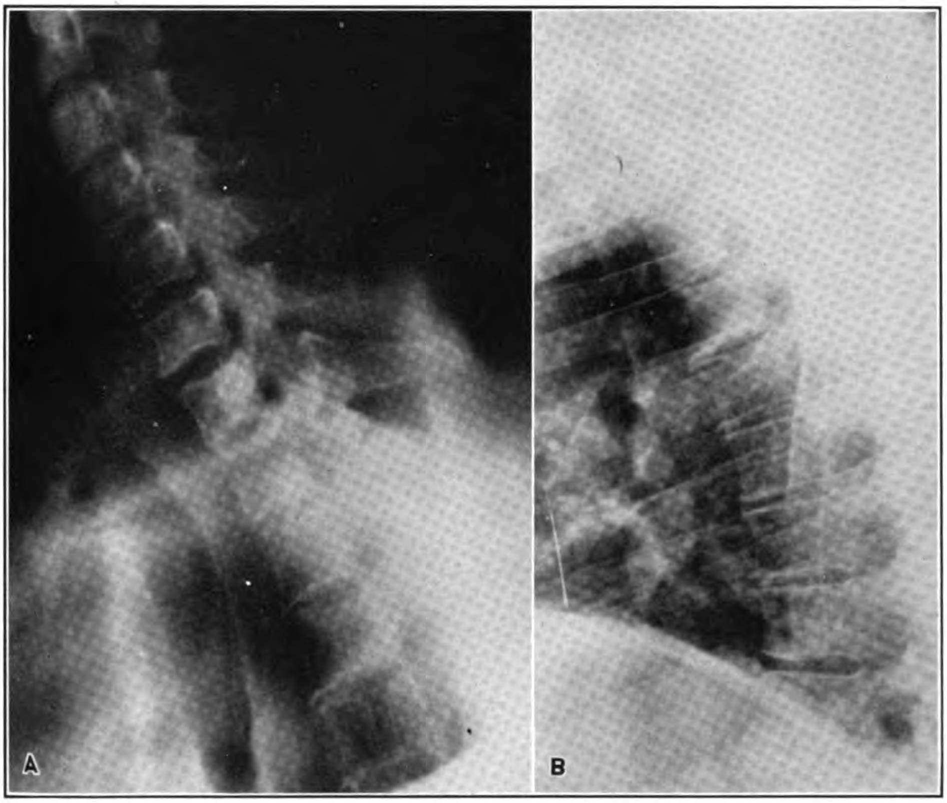


FIG. 190.—*A*, Lateral body section roentgenogram of the cervical and upper thoracic vertebræ. Metastases from carcinoma of the breast to the upper thoracic vertebræ anteriorly. *B*, Same case, lateral thoracic spine roentgenogram. Metastases to midthoracic vertebræ. Even though there is marked decalcification of the vertebral bodies no collapse is noted. The patient had no pain in these areas at the time the films were made.

In a description of vertebral metastases one must constantly keep in mind that metastases frequently are multiple. The extent of metastases to the individual vertebræ may be quite variable, and no absolutely constant pattern of involvement can be described for any specific type of neoplastic deposit.

Metastases to the neural arches, the pedicles and the transverse and spinous processes are quite common. Neoplastic deposits in the pedicles and laminæ are of particular importance because of their close relationship to the spinal canal and the probability of pressure against the spinal cord or cauda equina by direct extension. When such is suspected stereoroentgenograms of the spine may be helpful in studying the configuration of the pedicles. The initial change may be nothing more than decalcification of a

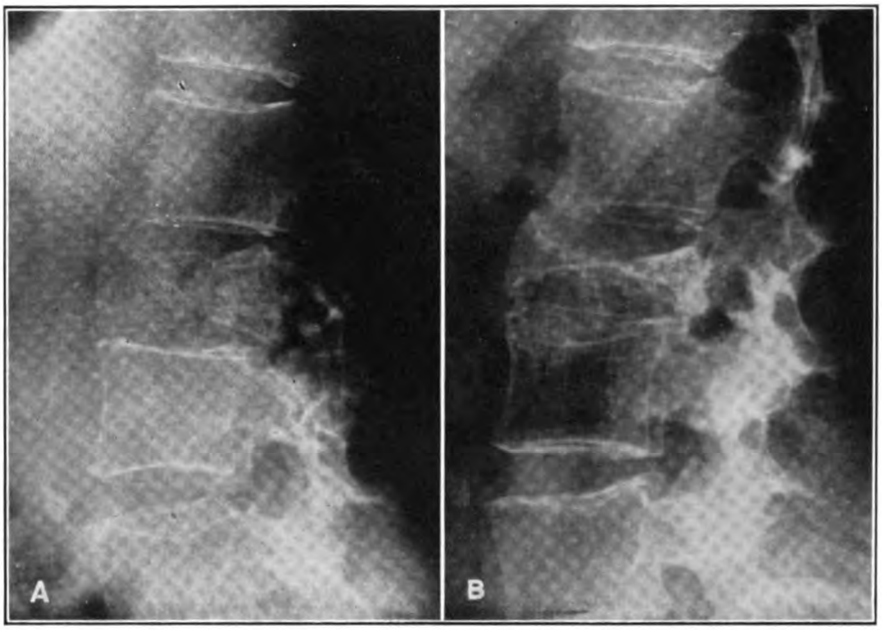


FIG. 191.—*A*, Metastases to the third lumbar vertebra from a carcinoma of the breast. Note the flattening of the vertebral body, its bulging contours and reticulated appearance. *B*, Same case, eight months later. *X*-ray therapy during that time had resulted in amelioration of pain.

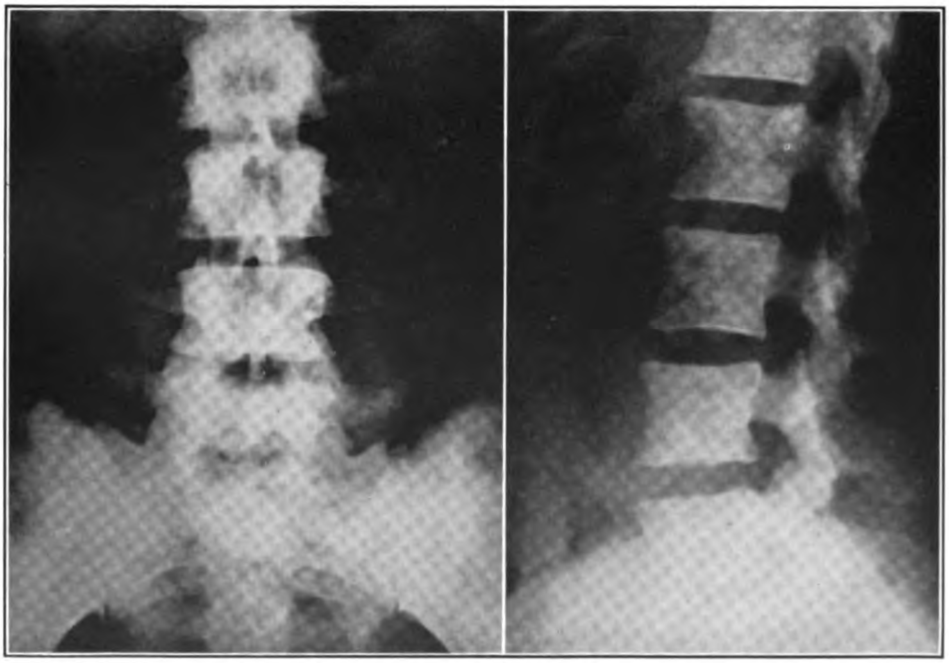


FIG. 192.—Anteroposterior and lateral roentgenograms of the lumbosacral spine. Predominantly sclerotic metastases from carcinoma of the breast.

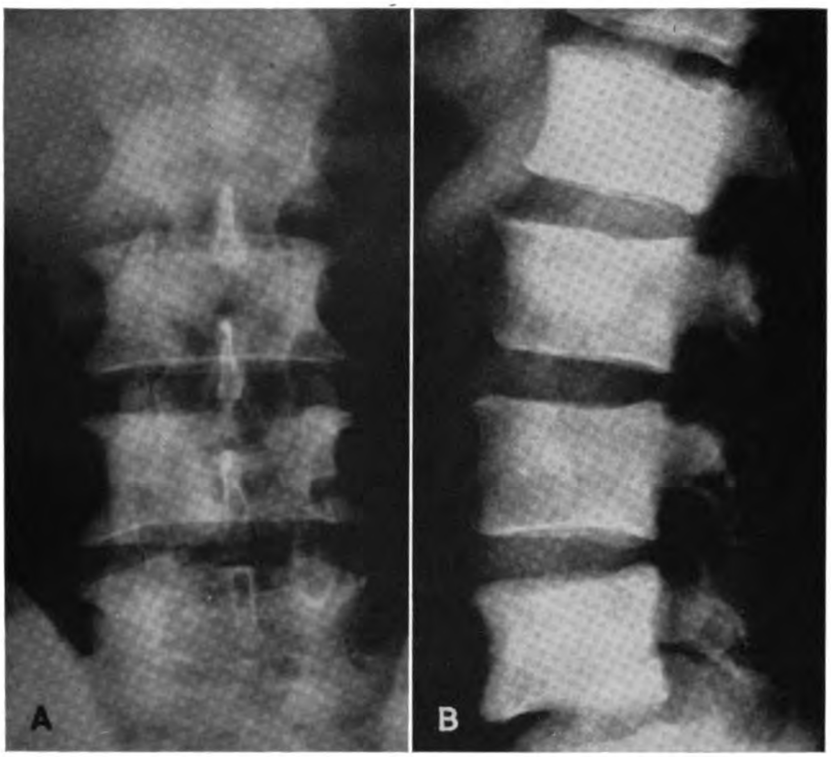


FIG. 193.—Mixed sclerotic and lytic metastases from a prostatic carcinoma. Note the destruction of the left lateral aspect of the fourth lumbar vertebral body and pedicle. *B*, The sclerotic element is better seen on the lateral projection.

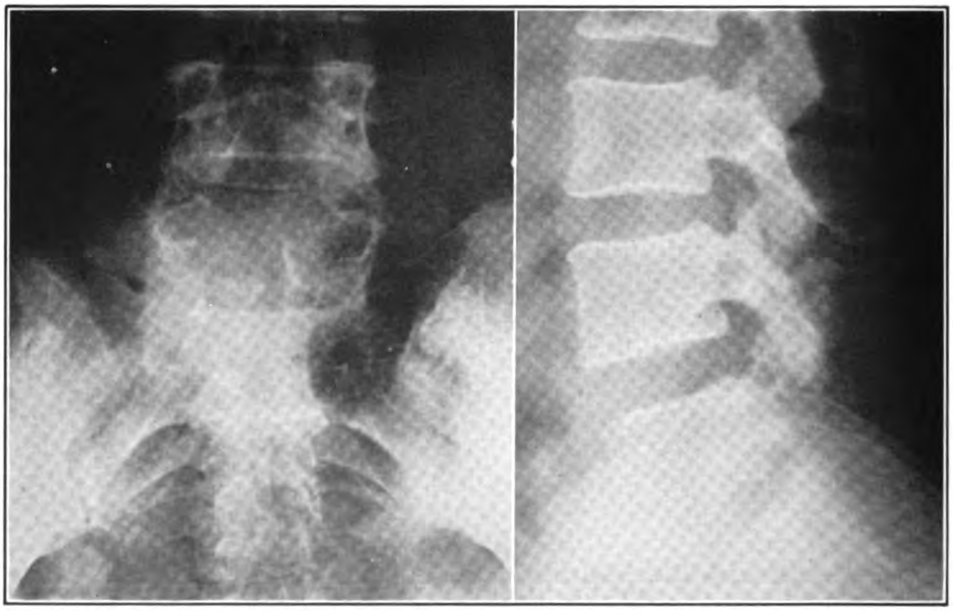


FIG. 194.—Lytic metastases to the neural arch, including the spinous process, of the fifth lumbar vertebra from a carcinoma of the lung.

single pedicle, with no bony reaction, and is easily overlooked. In more advanced cases a destructive process may extend into the lamina. Involvement of the transverse processes usually can be easily identified because these areas are not covered by adjacent bone. Destructive changes in the spinous processes may sometimes be identified on the anteroposterior view by the absence of the shadow of the spinous process across the vertebral body. However, in such instances it is better to obtain lateral soft tissue studies coning down on the suspected area.

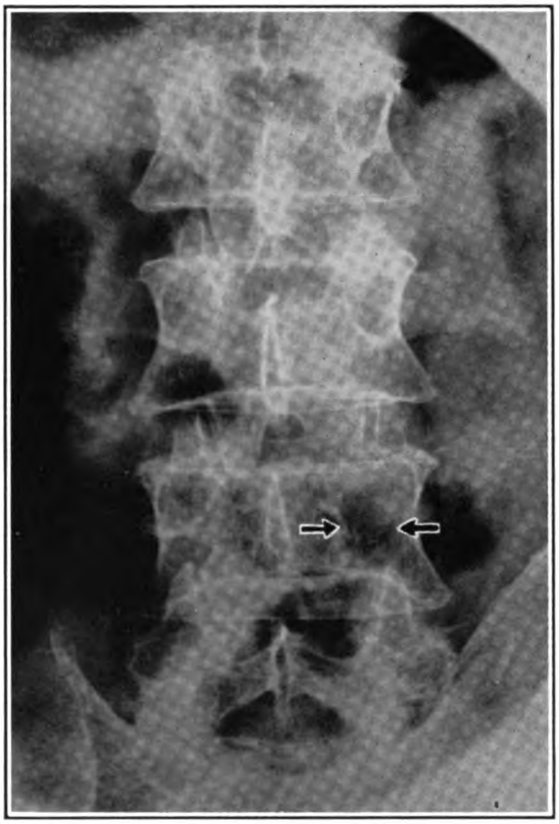


FIG. 195.—Lytic metastases to the left pedicle of the fourth lumbar vertebra extending to the inferior articular process (arrows) from a carcinoma of the kidney.

The effect of intraspinal metastatic deposits as a cause of pain and other neurologic symptoms is well recognized. As a rule, these deposits are associated with involvement of the adjacent vertebral body or neural arch, and produce cord compression and spinal canal block. Occasionally one encounters an epidural deposit without bone destruction. In others, the osseous involvement may be minimal, and situated at quite a distance from the intraspinal metastasis. When such deposits are situated low in the spinal canal the clinical picture may simulate a herniated disc. This occurred in 9 of 11 cases described by Epstein (1952). In 4 of these, the plain roentgenograms made before myelography were considered to be normal, while in 2 others there was discussion as to whether a slight decrease in the density of the fifth lumbar or first sacral segment indicated metastatic involvement. In 4 cases a faint lytic lesion of the fifth lumbar or first sacral vertebra had been overlooked, only to be identified on myelograms and a later review of the preceding films. In 1 case the metastatic nature of a very faint decalcification of the body of the fifth lumbar vertebra in its posterosuperior aspect was confirmed by the coexistence of ischial metastases. Myelographic examination of this group of patients showed what appeared to be a fairly characteristic defect, consisting of a deviation and tapering of the caudal sac contralateral to the side of the tumor deposit. In these cases the caudal sac assumed the appearance of an inverted inclined cone

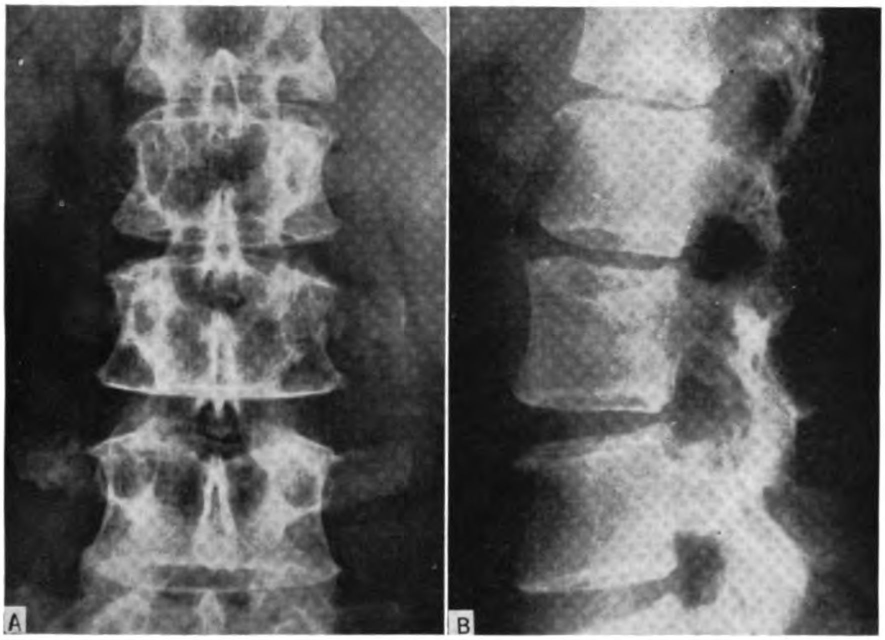


FIG. 196.—Multiple lytic metastases to the transverse processes of the lumbar vertebræ. The right pedicle of the second lumbar vertebra and the left pedicle of the third are destroyed. There is diffuse lytic change in the vertebral bodies, with partial collapse of the superior aspect of the third lumbar vertebra (*B*).

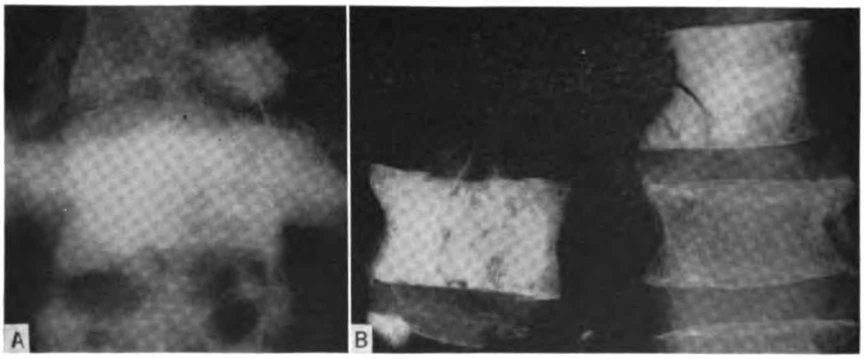


FIG. 197.—Intensely sclerotic vertebral metastasis from synovioma of a sacral nerve root, verified at necropsy.

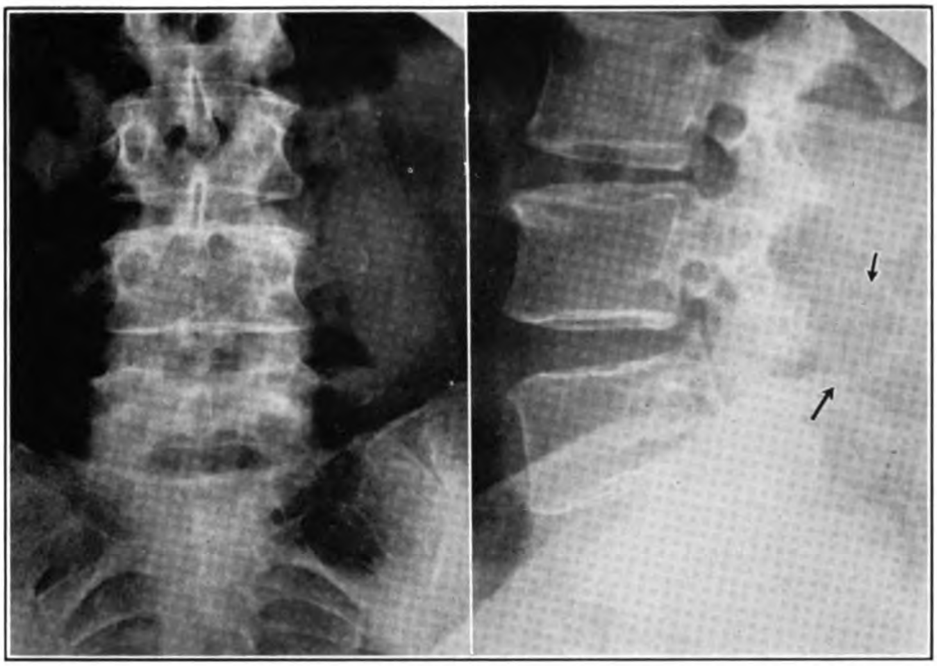


FIG. 198.—Anteroposterior and lateral lumbar spine roentgenograms demonstrating lytic metastases to the spinous process and neural arch of the fourth lumbar vertebra (arrows) from a melanoma of the eye.

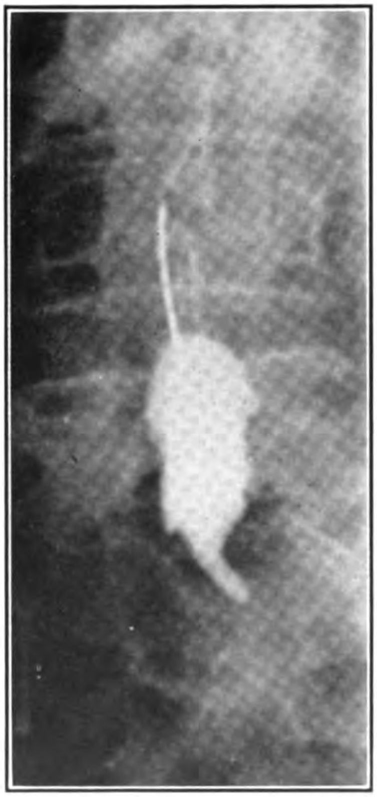


FIG. 199.—Displacement of the caudal sac from right to left by epidural metastases from a carcinoma of the lung.

of varying height deviated away from the lesion, while the ipsilateral axillary pouch at the lumbosacral interspace was blunted or obliterated. The axillary pouch on the opposite side was either pressed against the pantopaque column, or was unidentifiable. This defect was noted in 7 cases, but in the others the deformity could not be definitely differentiated from that which might be caused by a protruded nucleus pulposus. At operation it was noted that the deposits of malignant tissue were irregular, nodular in shape and varied in thickness from a few millimeters to over 1.5 cm. The nerve roots were deflected by the metastases, and in 2 cases encrustations of tumor tissue about the involved nerve roots were noted. Interestingly enough, no invasion of the dura itself was identified. In 9 cases invasion of bone a short distance from the epidural metastases was found at operation but no visible connections were noted between the osseous malignancies and the epidural deposits.

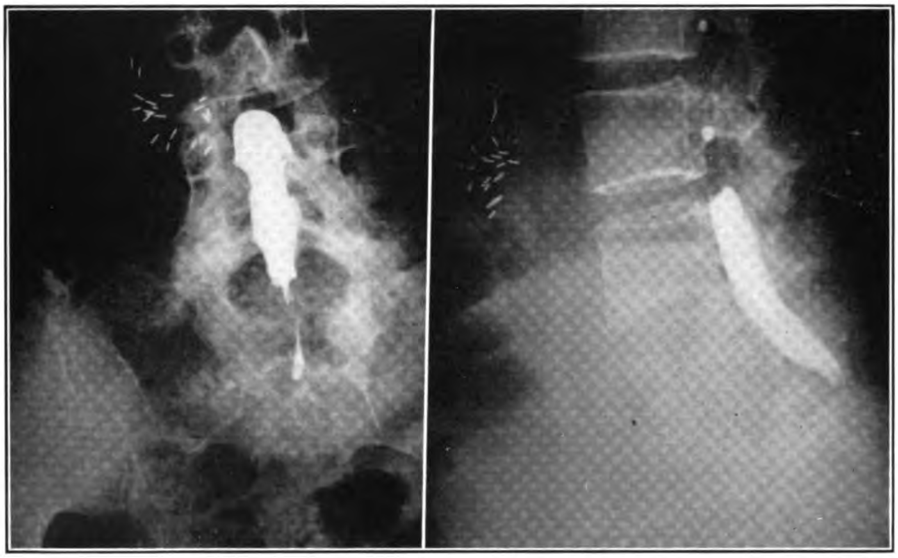


FIG. 200.—Lytic metastases to the right superior sacrum. Epidural metastases constrict the caudal sac on either side at the lumbosacral interspace and push the thecal sac dorsally as seen on the lateral myelogram. The primary lesion was a carcinoma of the pancreas.

Metastatic deposits high in the lumbar or in the cervico-dorsal spinal canal may be associated with symptoms of spinal cord compression. In such instances destruction of bone points towards the involved area. However, myelographic examination in many of these individuals will show that the block may be somewhat distant from the visible bony destruction and at operation extension of neoplastic tissue into the spinal canal can be seen to account for this discrepancy. In our patients with metastases to the upper lumbar, thoracic or cervical spinal canal with myelograms the usual change was partial or a complete arrest of the passage of pantopaque. In a few instances the column was deviated contralaterally passing above the area of block through a thin lateral or anterior channel. In the great majority of these cases adjacent bony destruction was present but in several instances the bone appeared to be intact both as observed radiologically and as described at operation. However, since only the neural arches were investigated in these individuals one can make no statement as to what was going on in the vertebral body itself, even though the roentgenograms were normal.

Intraspinal metastatic deposits occasionally occur with lymphomas. Smith and Stenstrom (1948) noted this in 11 out of 308 cases of Hodgkin's disease. Symptoms of spinal cord block were present in 3 cases. I have observed 6 similar cases. One patient, a 17-year-old boy with a verified lymphoblastoma, converted into an acute lymphatic leukemia. During this interval he developed symptoms of spinal cord compression at about the level of the tenth thoracic vertebra. Myelographic examination disclosed a complete block at this site, and laminectomy revealed a cuff of tumorous tissue extending cephalad for about 5 centimeters. On histologic examination this tissue was found to be composed of leukemic cells. The patient did well after operation and postoperative *x*-ray therapy. He expired about five months later, and necropsy disclosed extensive metastases from his

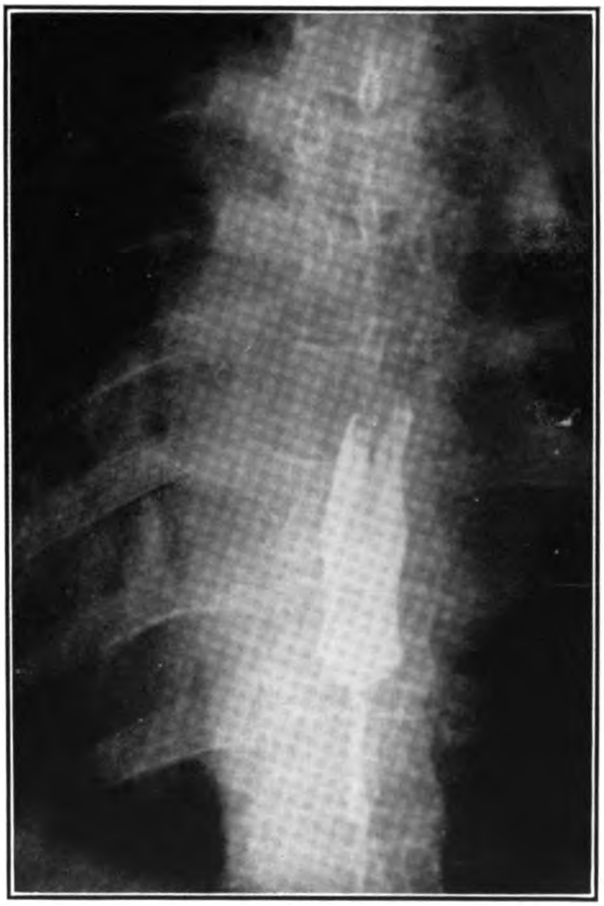


FIG. 201.—Metastases from breast carcinoma to the midthoracic vertebræ causing epidural spinal canal block. Both the fifth and sixth thoracic vertebræ are affected. The pantopaque block is at the inferior aspect of the sixth thoracic vertebra, which has destroyed pedicles as well as neural arch and vertebral body involvement.

lymphoblastoma. Apparently the leukemic transformation had receded back to the original condition. No evidence of recurrence of the spinal canal lesion was found. At operation the bits of bone removed for laminectomy disclosed deposits of leukemic cells, while the necropsy report indicated the existence of bony involvement with lymphoblastoma. In 2 other individuals with known lymphomas a preoperative diagnosis of spinal canal block by lymphomatous tissue was made because of the development of spinal cord compression. At operation, 1 of these patients was found to have an ependymoblastoma, and the other had a metastasis from an unsuspected carcinoma of the lung. The obvious moral is that one cannot assume with complete certainty the histologic nature of a lesion because of a coexistent neoplasm elsewhere.

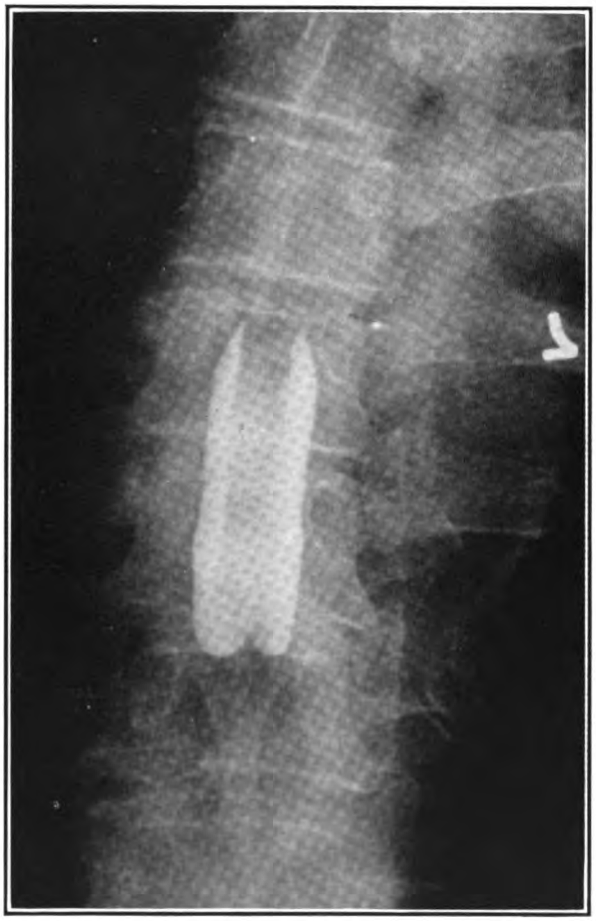


FIG. 202.—Intraspinal epidural metastases, midthoracic region from a carcinoma of the breast. The plain film roentgenograms were normal, and no osseous destruction was observed at operation.

The extension of intracranial tumors into the spinal canal by way of the cerebrospinal fluid circulation was first mentioned by Cairns and Russel (1931), who observed gross evidence of spinal metastases in more than one-third of their cases. More than half were medulloblastomas. The others were classified as glioblastoma multiforme, astrocytoma, ependymoma,

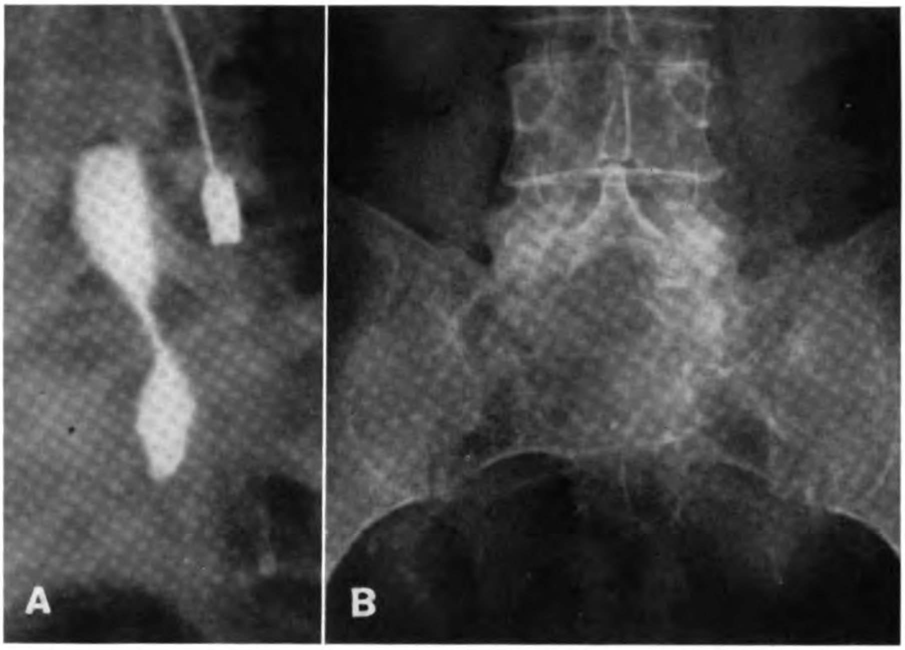


FIG. 203.—Epidural metastasis from "benign" metastasizing thyroid deforming the thecal sac. The clinical diagnosis had been a herniated disc. The thyroid gland was not enlarged, and plain spine films were normal. At operation the bone appeared normal. Relief of symptoms followed operation and *x*-ray therapy. Four years later (*B*) pain recurred and osseous metastases were found in the right side of the sacrum extending towards the midline.

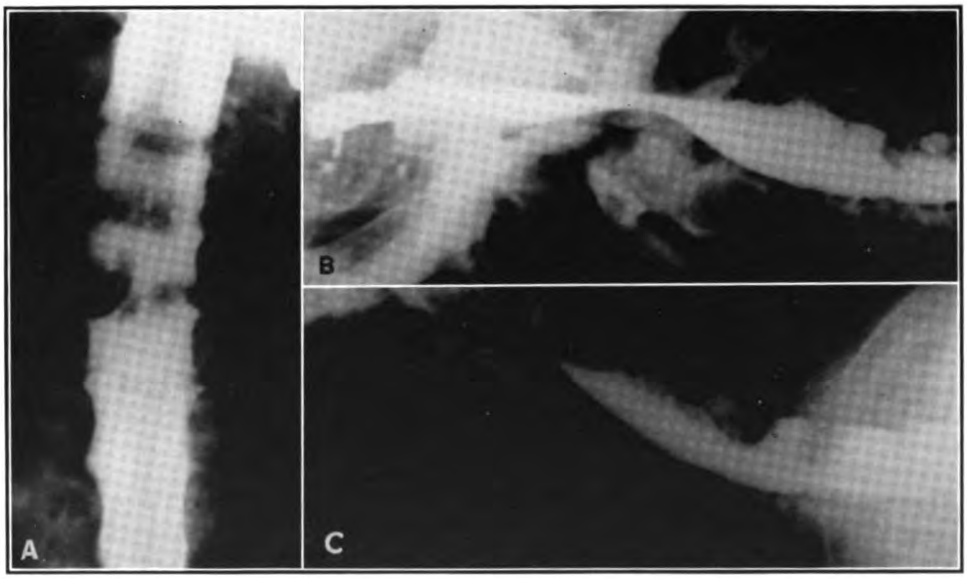


FIG. 204.—Metastatic tumor destroying the dorsal root ganglion at the right sixth cervical segment in a 58-year-old woman with a clinical syndrome of a cervical herniated disc. No evidence of bone involvement was found on preliminary cervical spinal roentgenograms, but at operation the adjacent lamina was found to be involved. Above the lateral defect are several ridge deformities (*A*). On transcervical studies (*B* and *C*) the transverse deformities were observed to be more pronounced with the neck in flexion, and were incident to thickening of the yellow ligaments. They were considered to be of some importance in aggravating the effect of the lateral lesion, which had been diagnosed as a herniated disc or ridge. Verified at operation. Patient had had a mastectomy five years before.

neuro-epithelioma, and one was identified as a glioma of the third ventricle. Half originated above the tentorium and half below. Wolf (1941) mentioned that spinal metastases from brain tumors constituted 4 per cent of all spinal cord neoplasmas. A case of cerebellar medulloblastoma in which metastases were carried by silver clips from the brain to the spinal cord was described by Halpern (1942). Implants within the dural sac in the sacral region from an ependymoma in the third and fourth cerebral ventricle was reported by Chusid (1948), who described further extension of the tumor into the epidural tissue, probably along the perineural spaces. This author cited 12 similar cases from the literature. The myelographic demonstration of spinal cord metastases from brain tumors was described by Wood, Taveras and Pool (1953). They observed 2 types of glioma metastases. One form was discrete nodular implants along the spinal cord and nerve roots, more often on the dorsal aspect. These varied in size from microscopic implants to nodules several centimeters in diameter, and occasionally invaded the parenchyma itself. In such cases the cord might be compressed or irregularly swollen. The second and relatively infrequent type of pathologic change was thickening and opacity of the leptomeninges secondary to a diffuse type involvement. In such cases the tumor tissue varied in thickness from a thin film to a sheet several millimeters thick. Microscopically, the tumor may resemble the parent neoplasm. According to Wolf (1941) those which invade the meninges may differ somewhat because of fibrosis of the neoplasm, giving a sarcomatous appearance. Wood, Taveras and Pool reported 4 patients, 2 with glioblastoma multiforme and 2 with cerebellar tumors (medulloblastoma and hemangioblastoma). In 3 cases the metastases involved the spinal cord primarily, while in the

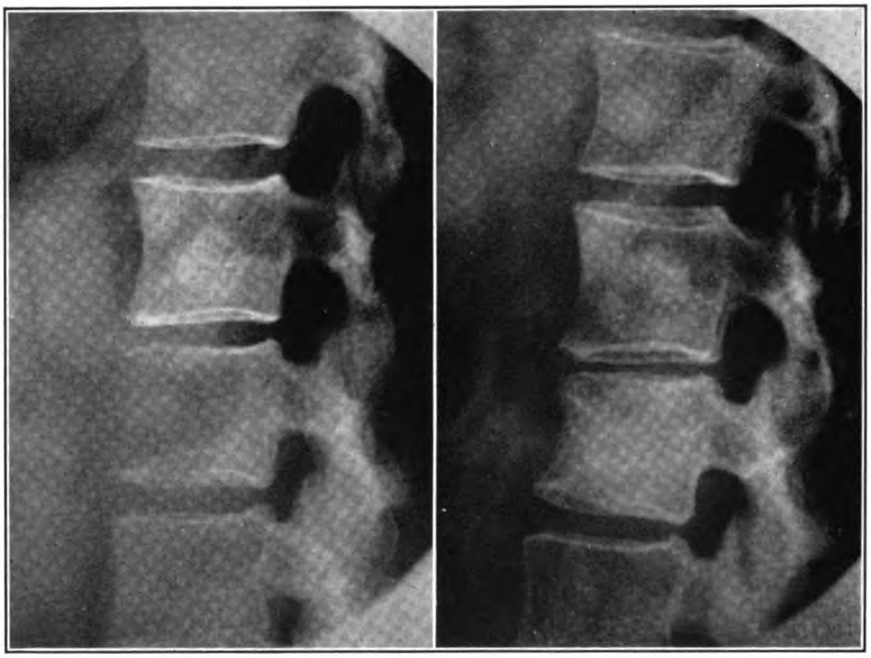


FIG. 205.—Lytic metastases from carcinoma of the breast to the second lumbar vertebra. Restoration of bone after *x*-ray therapy.

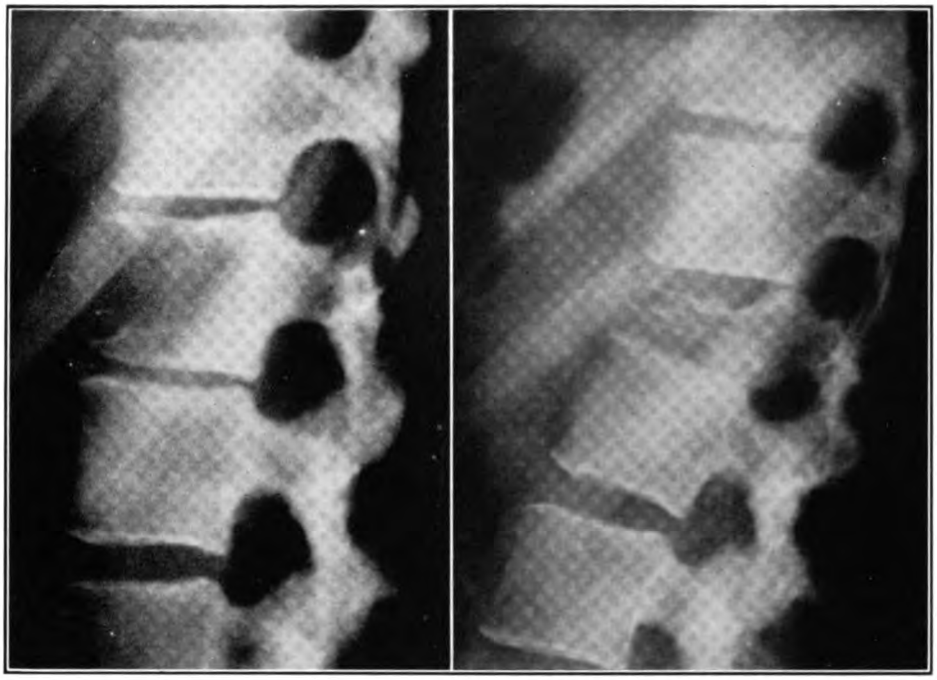


FIG 206.—Lytic metastases to the twelfth thoracic vertebra from carcinoma of the cervix. Within four months the vertebra collapsed so that a vertebra plana resulted, with narrowing of the intervertebral foramina between the twelfth thoracic and first lumbar vertebra. The intervertebral discs were undisturbed.

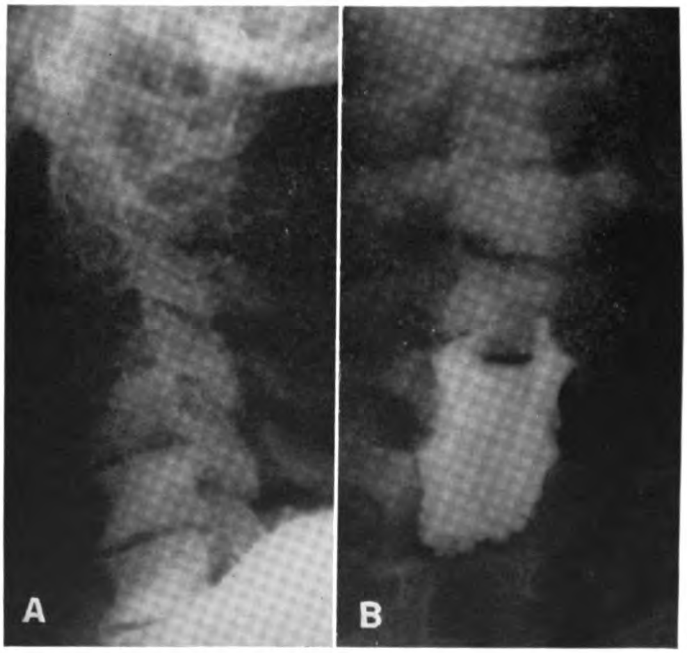


FIG. 207.—Cervical sclerotic metastases from a prostatic carcinoma in a 58-year-old man with pain in both arms and shoulders for 4 weeks, with partial paralysis of both upper extremities. Complete pantopaque block at the level of the sixth interspace. Epidural extension of tumor at operation, with relief of symptoms after decompression.

fourth only the cauda equina was involved. Myelograms of these patients revealed multiple oval or rounded constant filling defects which were partially or completely obstructive. In 1 case a myelogram before and after irradiation showed disappearance of the nodules. The plain roentgenograms in these patients were all normal.

The effectiveness of *x*-ray therapy in restoring bone in lytic metastases of the vertebral column is well known and requires no further comment here. Mention must be made of the great usefulness of hormonal therapy in producing recalcification of vertebral bodies.

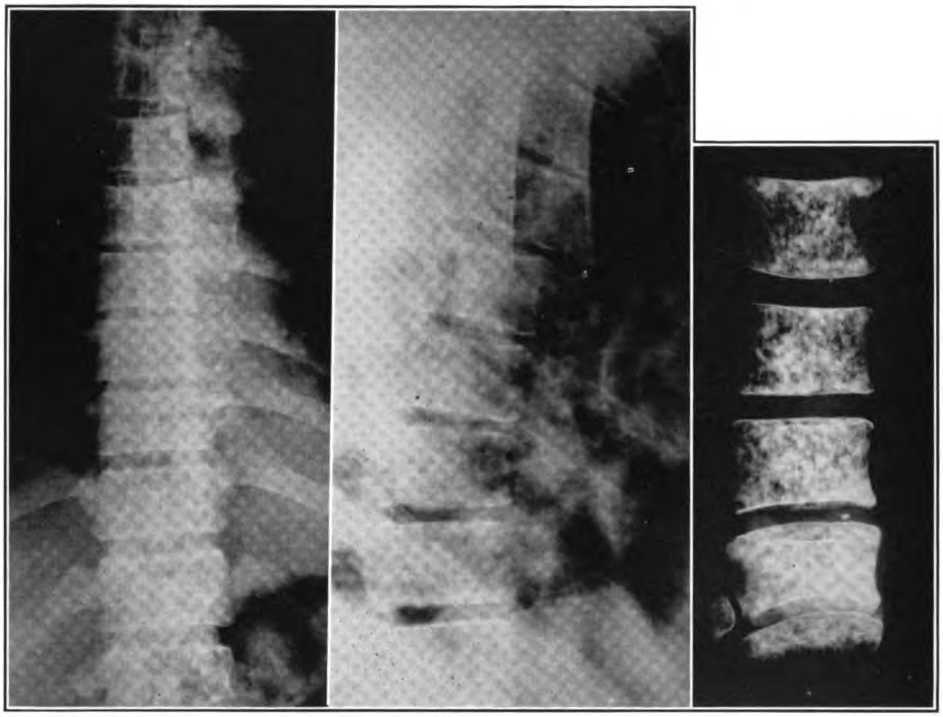


FIG. 208.—Spotty osteoblastic metastases to the spine from an unsuspected carcinoma of the stomach, with necropsy verification.

It should be reiterated that neoplasms almost anywhere can spread to the vertebræ. Among these may be mentioned tumors of the heart (Woll and Vickery, 1947), the liver (Auerbach and Trubowitz, 1950), the uterus (Robbins, 1943), the cervix, the lip (Tyler, 1942), the bladder (Freid, 1946), the ovaries (Pendergrass and Selman, 1946), the testes (Lafferty and Pendergrass, 1950), melanomas (Russo, 1947), and various situations along the gastrointestinal tract (Aufses, 1930; Stein, 1940; Bertin, 1944; Abrams, 1950), the pancreas (Scott, 1952), the adrenals (Cross and Pace, 1950), and carcinoids (Toomey and Felson, 1960).

Special mention is made of skeletal metastases from gastrointestinal malignancies. With these may occur a form of osteosclerosis within the medullary canals of the long bones as well as of the vertebral bodies, producing diffuse and spotty areas of increased density. This may be associated with secondary anemia. Two cases observed by us had numerous small nodular shadows of increased density within the vertebral bodies as well as in the medullary canals of the long bones.

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MYELOMA

Myelomas are fairly common tumors which arise from a single cell type in the bone marrow. Depending upon the predominant type of marrow cell involved, four main varieties are recognized—the plasma cell myeloma, myelocytic myeloma, erythroblastic myeloma and lymphocytic myeloma. The plasma cell myeloma is the type generally encountered. This classification is of greater pathologic than clinical significance, inasmuch as the gross pathologic changes and clinical course are practically identical in all.

In the primary stage of the disease involvement is chiefly in the distribution of the red bone marrow. Later, the viscera such as the liver, spleen, kidneys, lymph nodes and occasionally the lungs may be affected. The bones most frequently involved are the spine, the ribs, the pelvis, the skull and the long bones approximately in the order named.

The usual clinical picture is one in which pain predominates. The initial complaints are often referred to the back or the chest, and may be mild until exaggerated by a pathologic fracture following mild trauma. Occasionally the initial complaints are produced by extension of the tumor into the adjacent soft tissues producing neuritic, pleural or abdominal pain. Other fairly common complaints are progressive weakness and anemia. In the late stages of the disease various manifestations of visceral disturbances may occur. These involve the gastrointestinal, the genitourinary and particularly the nervous system. Neurological complications are due to pressure on the neural structures or their blood supplies. This results from tumor perforation of the vertebra and impingement on the spinal canal or the peripheral nerves, or by compression of the spinal cord or nerve roots by fractured vertebræ. Primary epidural myeloma unaccompanied by bony disease is relatively rare, and usually presents the clinical and myelographic appearances of an intraspinal tumor. More often the myelomatous osseous destruction produces rarefaction of one or more vertebræ, including the pedicles and neural arches, and the initial complaints are referable to the bony changes. With collapse of bone and extension of the myeloma into the spinal canal pressure on the cord and cauda equina are followed by the usual sequelæ. Occasionally a peripheral neuritis of unknown pathogenesis occurs with multiple myeloma.

A clue to the diagnosis can be obtained from blood chemistry and electrophoretic studies, the chief changes encountered being hyperproteinemia with reversal of the albumin-globulin ratio. However, in this regard one should keep in mind that similiar changes may occur in a wide variety of diseases such as cirrhosis of the liver, chronic nephritis, osteoporosis secondary to hyperthyroidism, myxedema, carcinomatosis and other rarer complaints. The peripheral blood count as a rule is not abnormal. Extremely important evidence for the existence of myeloma can be obtained by examination of the bone marrow.

When a considerable degree of osteoporosis accompanies myeloma, disturbances in the blood calcium and phosphorus content appear without depression of the blood phosphorus, and with normal serum alkaline phosphatase content.

Discussion has arisen as to whether myeloma may exist as a single isolated lesion. Cutler, Buschke and Cantril (1936) referred to two types of solitary myeloma, one in which the lesion was first solitary and later became generalized, and a second type in which the lesion was completely benign in nature, remaining localized throughout its entire course. From a review of the literature, Raven and Willis (1949) concluded that there were 18 acceptable cases of solitary plasmacytoma of the spine. This occurred more often in males. Two forms were described, one with a marked destruction of bone, the process being well demarcated and sharply defined, while the second was accompanied by a cystic and trabeculated appearance of bone, with irregular and thickened trabeculæ situated in the medulla and expanding the cortex.

Paul and Pohle (1940) believed that solitary myeloma of bone occurred as an unusual, but not rare disease. In a review of 45 cases, they described two forms, one simulating a giant cell tumor, the other an osteolytic type more frequently seen in the spine, and usually limited to a single vertebra. In some cases contiguous segments were affected. No proliferative bone changes were noted. The affected vertebral bodies frequently were flattened or presented lateral or anterior wedging and in some instances the vertebra practically vanished. The presence of paravertebral soft tissue masses in connection with multiple myeloma simulating abscesses was mentioned, and these authors believed that spinal myeloma might be confused with tuberculosis, particularly if the paravertebral soft tissue shadow was seen. In a review of 61 cases of solitary myeloma, the disease was found most often in the spine (Gootnick, 1945).

Christopherson and Miller (1950) also considered solitary plasma cell myeloma of bone a well-established entity, but believed it to be unrelated to multiple myeloma. They made a critical review of 94 cases recorded in the literature between 1897 and 1949, and presented 3 of their own. The lesions were considered solitary only if they had been followed for at least three years without dissemination, or if a necropsy had been performed and only a single lesion found. The vertebræ was found to be the most common site; the femurs and pelvis next in frequency. These authors mentioned two radiologic types, one similar to giant cell tumor, and the second purely destructive. In their opinion the lesion was histologically similar to plasma cell myeloma, but behaved more like a granuloma. The over-all picture was obscured by the large number of cases reported as solitary myelomas which later proved to be common multiple myeloma.

On the other hand, the opinion has been expressed that the so-called solitary myeloma was only expression of the early stage of the disease, originating in a single focus. Toth and Wintermantel (1943) reported a case originating in the pubic bone with subsequent generalization, and commented that the seemingly solitary myeloma was not in itself a specific condition. Aegerter and Robbins (1947), and Bayrd and Heck (1947), believed all myelomas to be multiple in nature, even though only a single bone might be involved initially and for a long time. White and Tillinghast (1950) also believed that the solitary form of myeloma was extremely rare and may be merely an early manifestation. Dalgaard and Dalgaard (1952) reported 3 lesions which first occurred in the vertebral bodies, but terminally other bones and organs were involved.

The presence of Bence-Jones proteinuria has been regarded by many as indicative of multiple myeloma. The incidence of urinary Bence-Jones protein with multiple myeloma has been variously estimated as from 10 to 85 per cent. As brought out by Geschickter and Copeland (1949) the reported incidence of such proteinuria would be greater if proper laboratory tests for its existence were made routinely. According to Lumb and Prossor (1948) Bence-Jones proteinuria is probably associated with total bone marrow involvement by tumor.

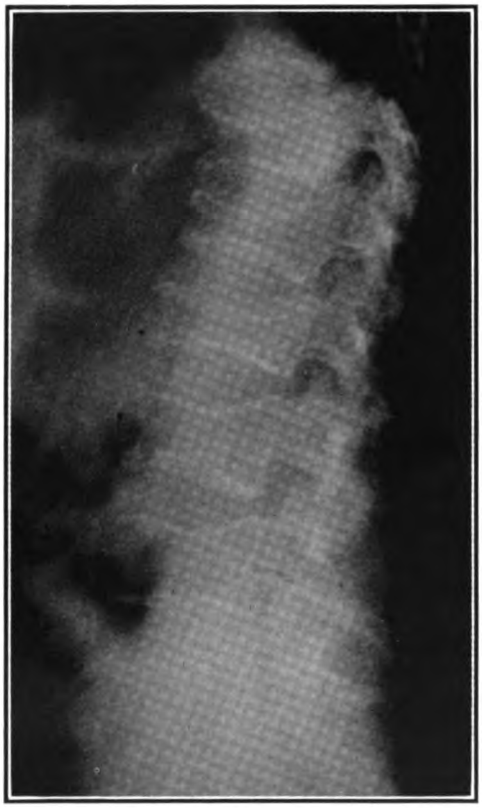

FIG. 209.—Pronounced osteoporosis with compressed vertebral bodies and widened intervertebral discs due to multiple myeloma.

The radiologic manifestations of myeloma include chiefly the punched-out, fairly sharply-demarcated rarefied areas observed particularly in the skull, the pelvis and long bones. Expanding lesions of the flat bones which give rise to a ballooned-out appearance of the ribs, pelvis and occasionally of the skull sometimes appear. These may be associated with perforation of the cortex and the appearance of subcutaneous soft tissue masses of myeloma. Occasionally this happens intrathoracically, and the lesion presents towards the pleural cavity as a semicircular, sharply-circumscribed nodular mass. In other instances the appearance of a myeloma is almost indistinguishable from that observed in metastatic malignancy of whatever origin.

The radiologic aspects of spinal involvement with myeloma present a varied picture of extreme interest. The usual effect of myeloma is an osteoporotic process. In its early stages, this is only slight, and occasionally associated with deformity of a vertebral body which may vary from lateral localized compression to a wedging or flattening of the involved vertebræ. Punched-out rarefied foci in the vertebræ, while less common, may occur. However, a diffuse form of osteoporosis interspersed in which are somewhat thickened spicules of bone is the more frequent lesion. Another fairly common manifestation of myeloma may be a diffuse osteoporosis of the entire vertebral column, almost indistinguishable from the osteoporosis associated with post-menopausal disturbances or idiopathic or senile osteoporosis. In these cases skull roentgenograms should be obtained. If parietal thinness can be demonstrated a benign osteoporotic process is more likely (Epstein, 1952). Extension of the myelomatous tissue beyond the confines of the vertebral bodies is rather infrequent, but may occur. Spread of the tumor across the intervertebral disc is uncommon, the extension probably following along the undersurfaces of the anterior and lateral spinal ligaments rather than penetrating the intervertebral cartilages.

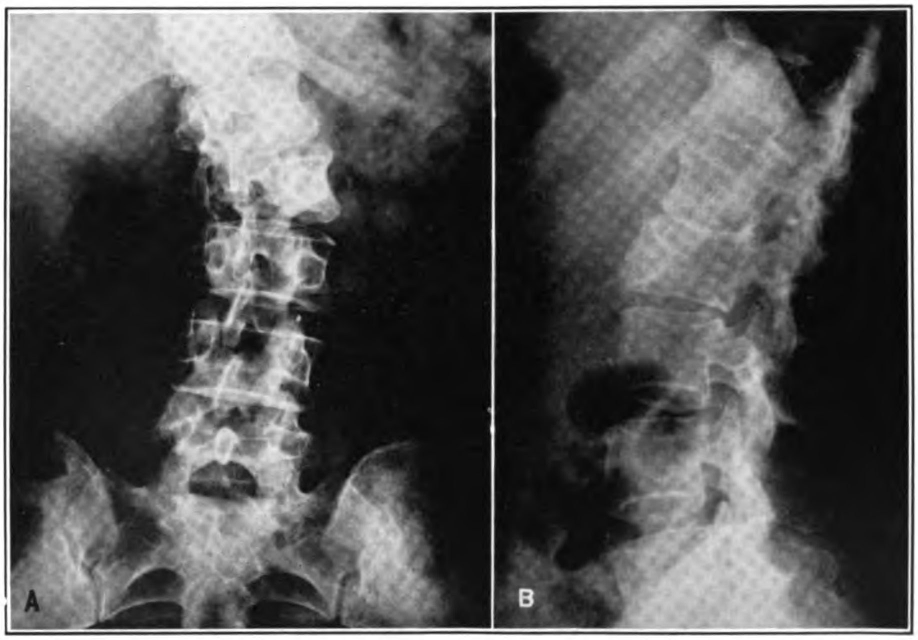

FIG. 210.—*A*, Collapse of the right lateral aspect of the body of the second lumbar vertebra due to multiple myeloma. *B*, Same case, lateral view. Bulging of the anterior aspect of the second lumbar vertebra is present.

With myelomatous involvement of a single vertebra, deformity of the body may be the first indication of the existence of a neoplastic lesion, and is indistinguishable from metastases. Careful scrutiny of the spine and skeleton in many of these cases may disclose other pathologic areas. The patient may present himself because of neurologic symptoms secondary to compression of the spinal cord. Myelographic examination will disclose either a partial or complete block. When the lesion is low in the spinal canal, the clinical picture mimics that of a herniated intervertebral disc. The myelogram may easily be interpreted as consistent with this condition. We have observed several such instances in which the bone lesion could not be accurately identified, and the true nature of the condition discovered only at operation. In these cases the myelographic defects were found to be due to nodular implants of myelomatous tissue in the epidural space displacing the pantopaque column contralaterally. In a few cases lesions at the lumbosacral articulation existed in which the plain films were so close to normal that the bone lesion could not be diagnosed even after postoperative scrutiny of the original roentgenograms. The myelograms in these individuals had presented evidence of a complete block at the lumbosacral interspace. Primary epidural myeloma cannot be diagnosed accurately without histologic proof of the absence of contiguous bone myeloma.

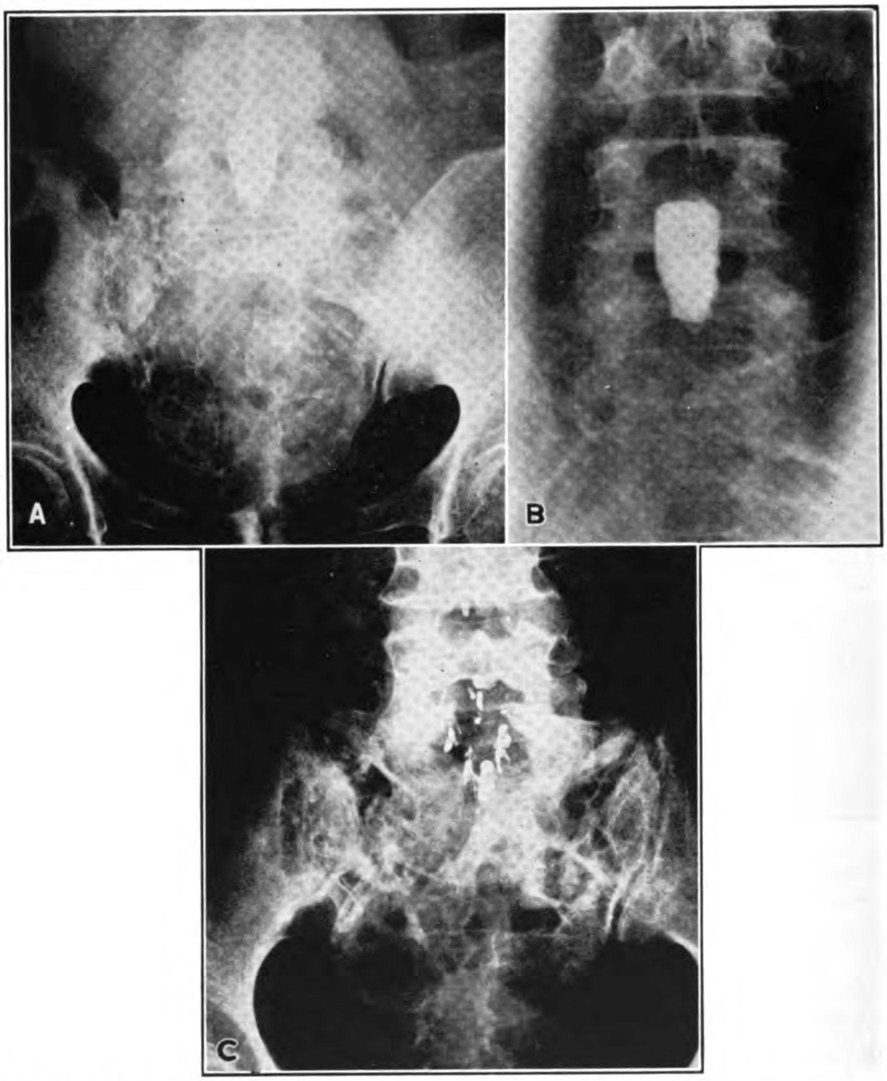

FIG. 211.—*A*, Myelomatous destruction of the right side of the sacrum. *B*, Same case. Complete block at the lumbosacral interspace noted on erect myelogram. *C*, Same case, with recalcification after deep *x*-ray therapy.

Occasionally one encounters a vertebral column in which a single vertebra

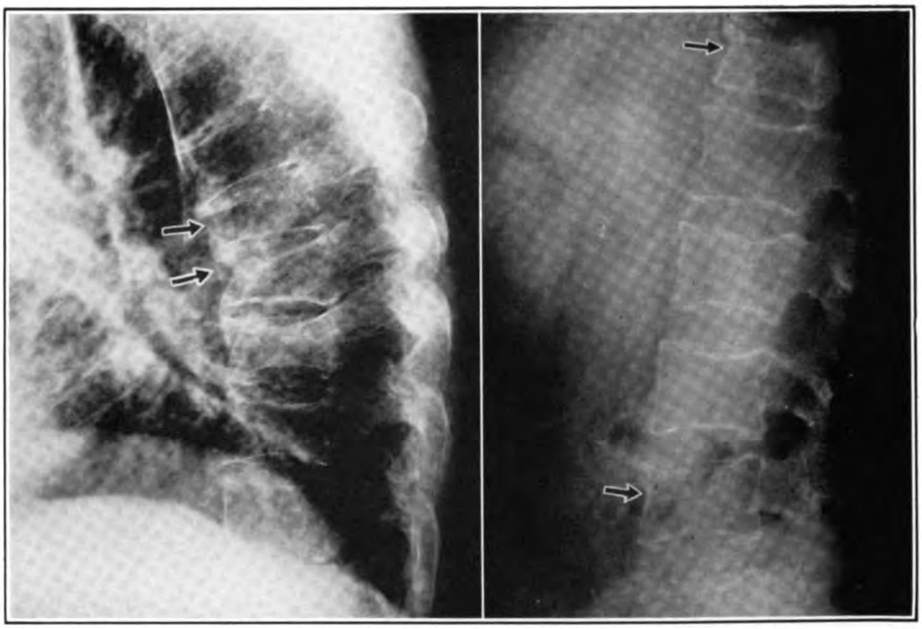

FIG. 212.—Osteoporosis and vertebral compressions due to multiple myeloma (arrows).

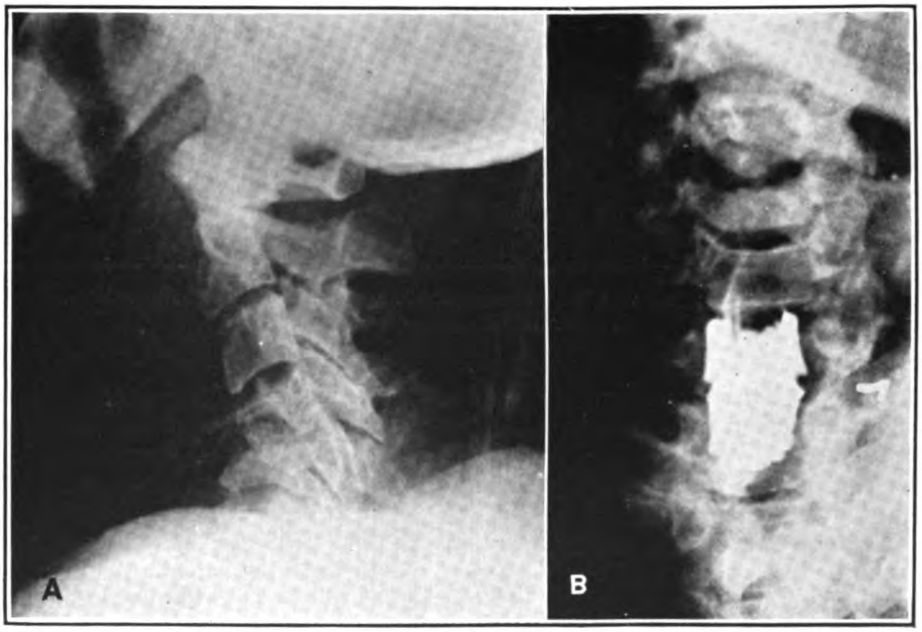

FIG. 213.—*A*, Myelomatous destruction of the fourth and fifth cervical vertebræ. *B*, Same case, with complete block demonstrated myelographically.

is partially collapsed and bulging beyond its normal confines. Associated with this there may be a number of other vertebræ which present a lesser degree of alteration of their configurations together with a rather reticular appearance of the medullary structures. Interspersed in these are occasional small areas of increased density secondary to partial collapse of cancellous tissue. In addition, compression fractures of the superior surfaces of one or several vertebral segments may be observed, particularly in the thoracolumbar region. These usually involve the anterosuperior aspect of the involved vertebral segments. Collapse of a lower cervical vertebra has been observed by us with a forward displacement of the neck associated with paraplegia. In this patient a complete block was noted myelographically. Narrowing of an intervertebral disc between two involved vertebræ occurs infrequently. We have observed several cases in which the vertebral body was completely flattened, with the intervertebral discs above and below relatively undisturbed.

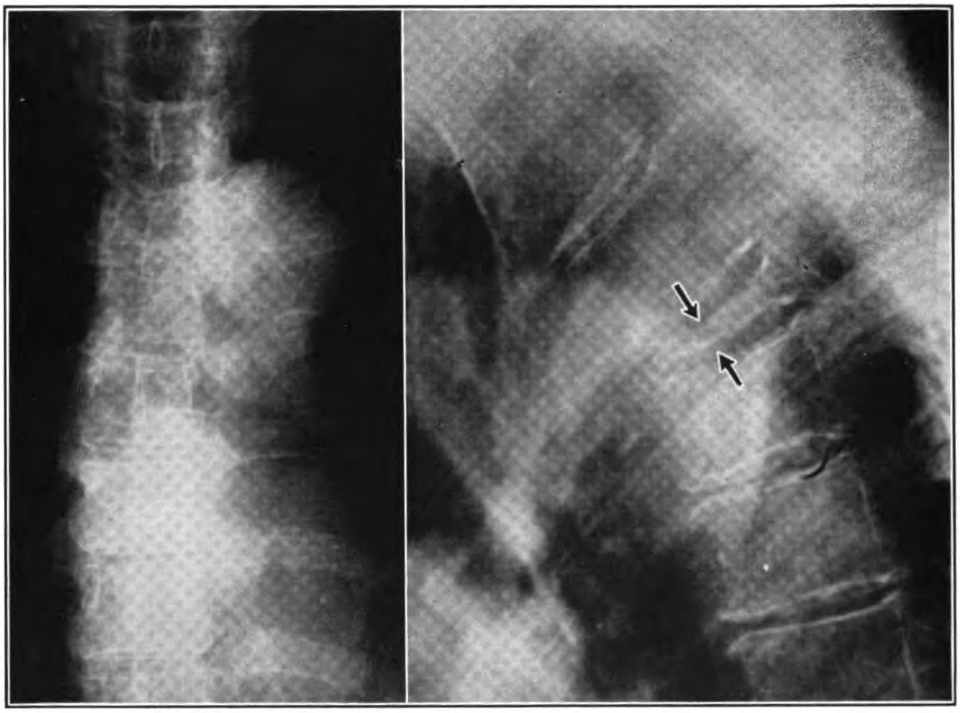

FIG. 214.—Collapse (vertebra plana) of the body of the sixth thoracic vertebra due to multiple myeloma (arrows).

Involvement of the transverse and spinous processes is somewhat uncommon, but occurs sufficiently often to warrant its consideration in the examination of any spine film. Of particular interest is the involvement of the neural arches with demineralization of a pedicle or lamina, particularly in the event of neurologic symptoms.

A rather rare form of myelomatous involvement of a vertebra is one in which the vertebral body becomes partially decalcified, and with dense linear bands traversing the area of decalcification. This form is referred to as resembling giant cell tumor. Three such cases appear in our series, in

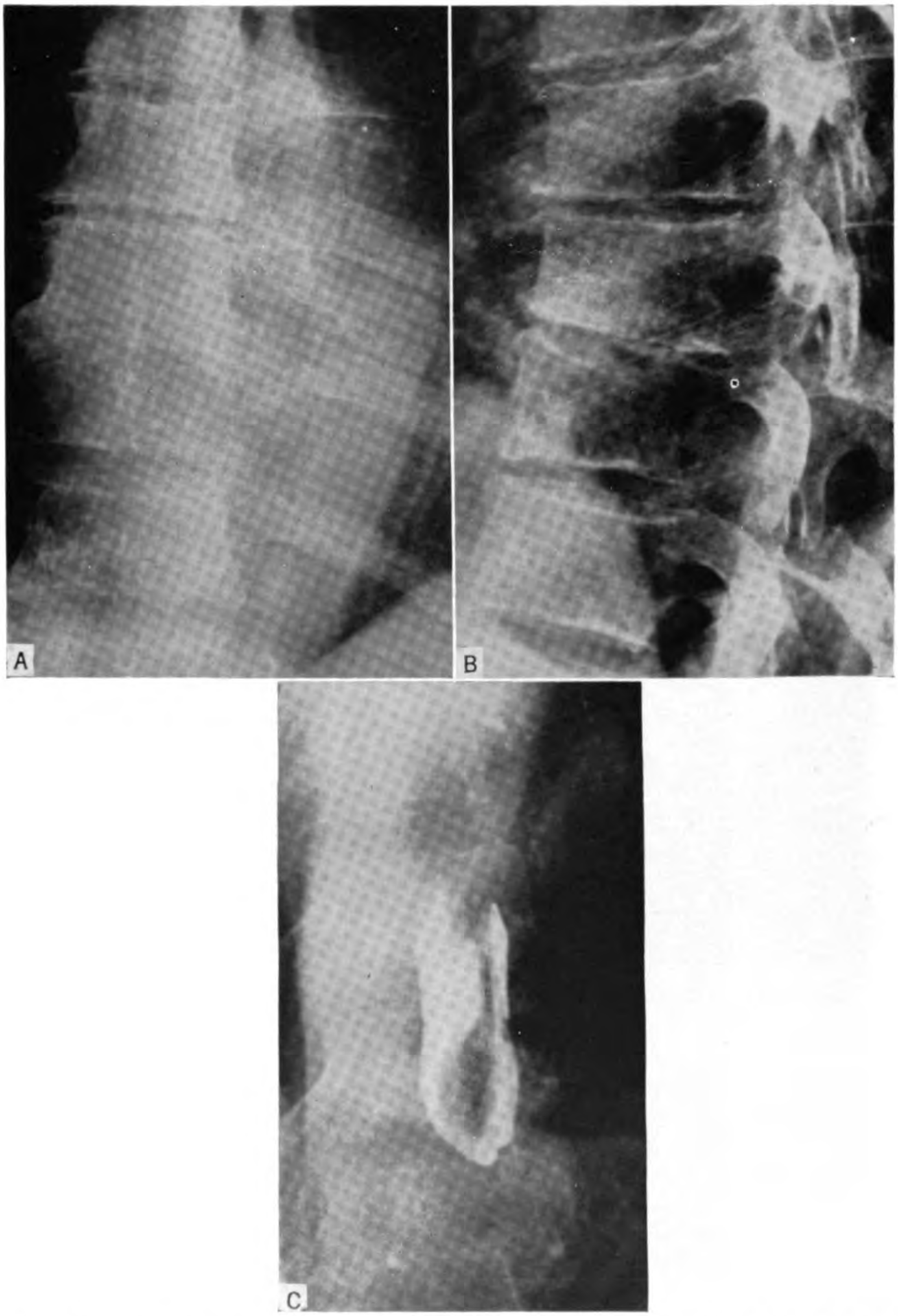

FIG. 215.—*A* and *B*, Myelomatous destruction of the posterior aspect of the ninth thoracic vertebra, its pedicles and neural arch, with extension of myelomatous tissue into the paravertebral area and destruction of left ninth rib head. The interspace between the eighth and ninth thoracic vertebra is narrowed. *C*, Complete block demonstrated on myelography.

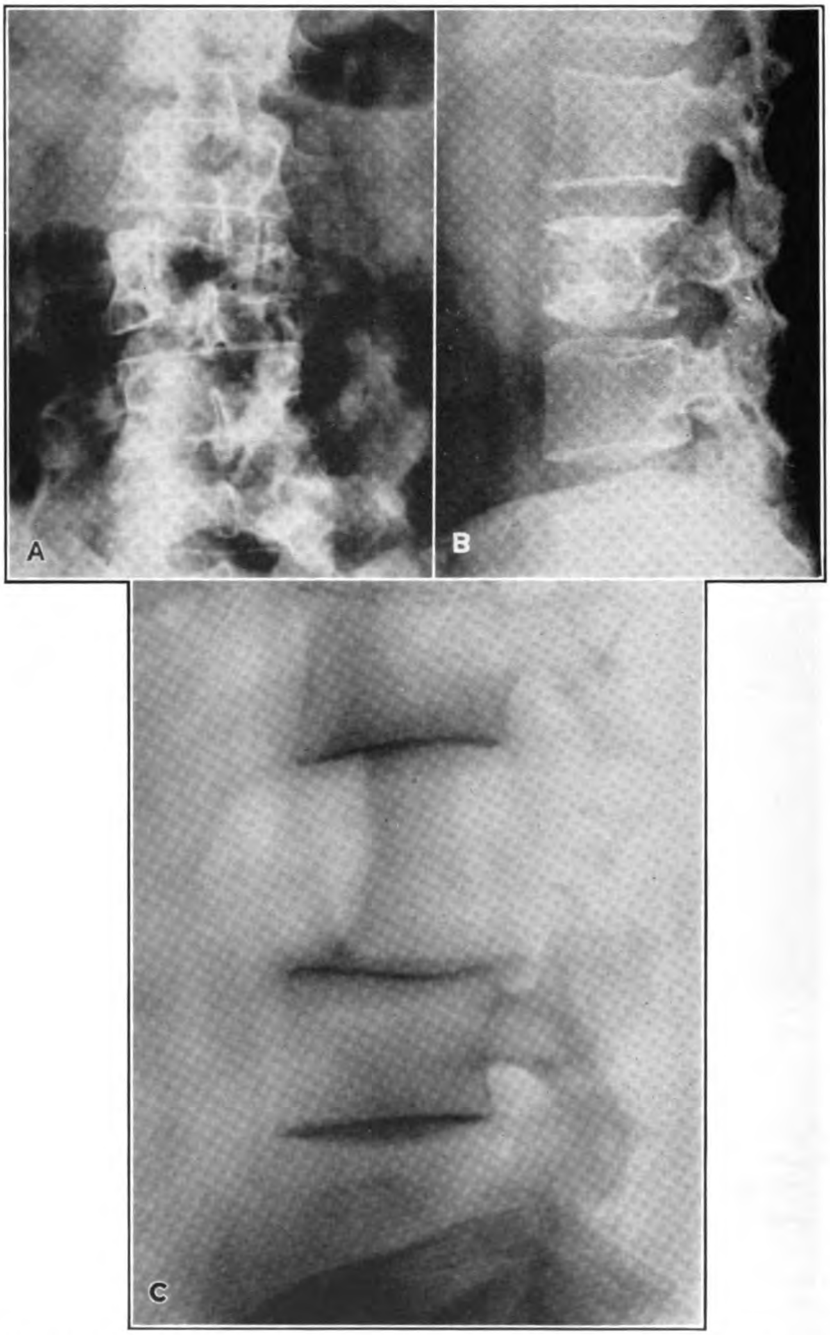

FIG. 216.—*A* and *B*, Collapse of the left lateral aspect of the body of the third lumbar vertebra. The reticular appearance as seen on the lateral view suggested the diagnosis of "giant cell tumor." *C*, Same case. Lateral lumbar spine roentgenogram made sixteen months later. There has been complete disappearance of the third lumbar vertebra. The fifth lumbar vertebra is practically gone, only a thin sliver of bone remaining visible. Diagnosis of multiple myeloma confirmed at necropsy.

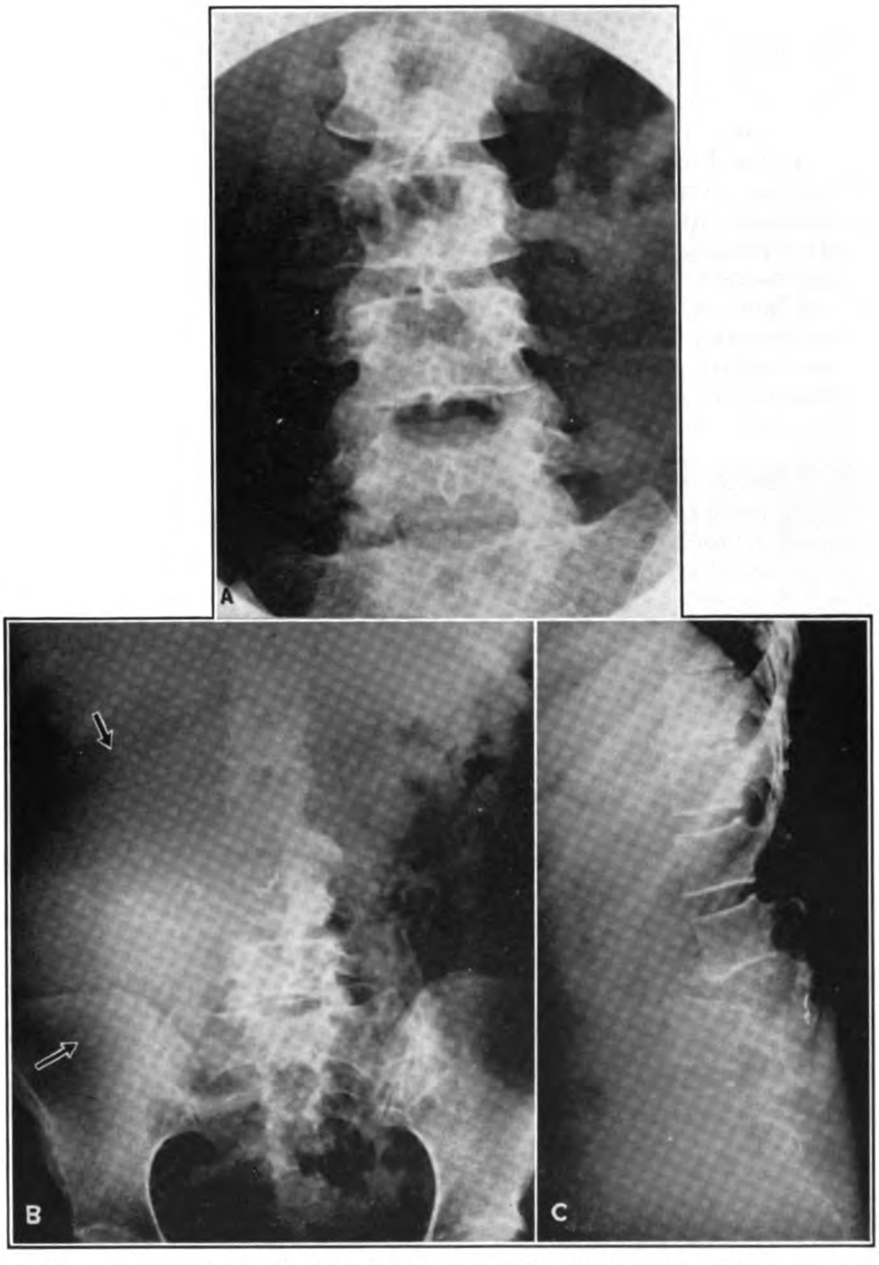

FIG. 217.—*A*, Expansion of the right lateral aspect of the body of the third lumbar vertebra, believed to be due to a benign bone tumor. *B*, Same case, two years later. The diagnosis of multiple myeloma had been established at operation done to relieve spinal canal block. A large paravertebral soft tissue mass practically filling the right side of the abdomen had appeared. Within this (arrows) was a reticular formation of new bone. *C*, Same case, lateral lumbar spine roentgenogram showing collapse of the third lumbar vertebra anteriorly. The posterior aspect is markedly involved as well, but remains somewhat higher.

each of which the diagnosis of giant cell tumor had been made. These patients had very little aside from a mild pain in the upper lumbar region. In 1 case followed over a two-year period, the true nature of the lesion manifested itself when the involved third lumbar vertebral body practically disappeared leaving a space equal to the height of the vertebral body plus the intervertebral disc above and beneath. In addition to the disappearance of this vertebra, the fifth lumbar vertebra practically vanished, leaving in its site only a thin, faint calcific shadow.

In another instance which came to necropsy the original lesion presented itself as a ballooning of the right half of the body of the third lumbar vertebra with decalcification of the right half of the body. Under subsequent observation, and as confirmed at necropsy three years later, the involved vertebral body was moderately demineralized. Associated with this was a tremendous mass projecting into the right side of the abdomen extending from the twelfth dorsal vertebra to the upper sacrum. Interspersed in this mass was a web-like amorphous calcification. At necropsy this was found to be myelomatous tissue, and extensive myeloma was found elsewhere in the body.

Osteosclerosis in myeloma is rare. Kohler and Laur (1950) cited 4 such instances among 179 cases in the literature and added 1 of their own. Engles, Smith and Krantz (1960) reported three more.

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ENDOTHELIOMA OF BONE (Ewing's Sarcoma)

Ewing (1940) described this neoplasm as an endothelial myeloma arising from the vascular endothelium in the bone marrow. Today it is generally accepted that the tumor more likely is derived from the mesenchymal connective tissue framework of the marrow. The basic cell type as described by Ewing was a small polyhedral cell with well defined margins and a small hyperchromatic nucleus. Jaffe attributed this to the effects of degeneration, and described cells with poorly delimited boundaries containing fairly large round or ovoid nuclei. The histologic differential diagnosis may be difficult, these cells being quite similar to those seen in skeletal metastases from neuroblastomas, reticulum cell sarcomas, lymphomas, multiple myelomas and other round cell tumors. Ewing's tumor arises most often in the long bones or the pelvis, and is mainly destructive, extending along the marrow cavity with little bony reaction. It occurs most often in young people and clinically simulates an infectious or granulomatous lesion with fever, leukocytosis and local pain. In differential diagnosis the possibilities include tuberculous or pyogenic osteomyelitis, eosinophilic granuloma and osteogenic sarcoma among the others mentioned above. While the tumor is radiosensitive, the prognosis is dismal. Very few patients survive for five

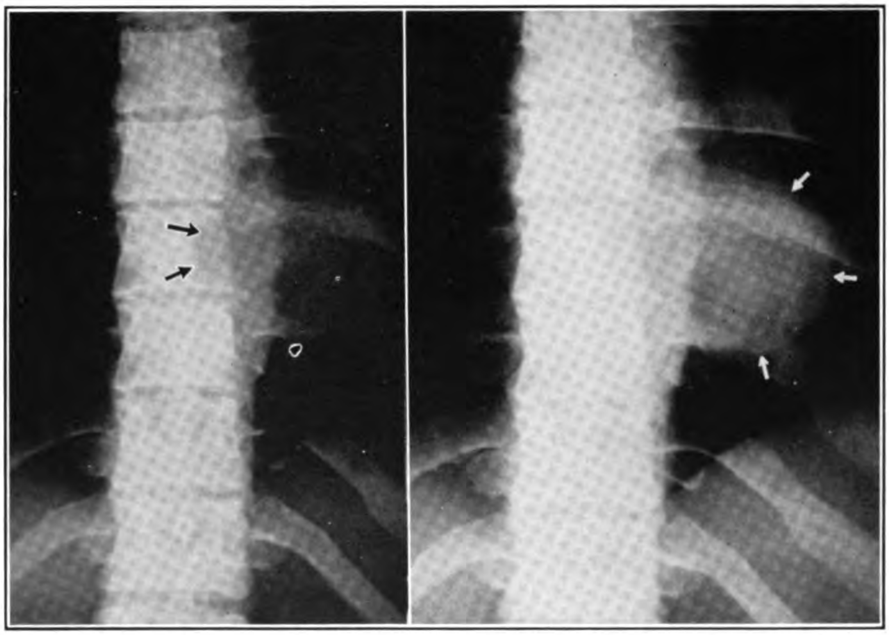

FIG. 218.—*A*, Ewing's tumor of the seventh thoracic vertebra. The left pedicle and the adjacent portion of the vertebral body are destroyed (arrows). *B*, Same case, soft tissue roentgenogram. Note the large paravertebral soft tissue mass extending to the left (arrows).

years. The tumor metastasizes widely, affecting the viscera, lungs and skeletal systems. When the vertebræ are involved the marrow spaces become extensively infiltrated, with areas of necrosis and perforations of the tumor under the paraspinal ligaments and into the epidural spaces.

The radiologic aspects of spinal involvement with Ewing's tumor are varied. Swenson (1943) reported a case in which destruction and wedging of the ninth dorsal vertebra had been interpreted as tuberculosis of the spine, and a spinal fusion had been done. In this case a paraspinal soft tissue mass was present. In a review of 37 cases Hamilton (1940) mentioned 2 with spinal involvement. One was a seventeen-year-old female with Ewing's tumor of the first and second lumbar vertebræ with slight kyphosis, tenderness and partial paraplegia, in whom treatment consisted of laminectomy followed by *x*-ray therapy. This patient was living seven and one-half years later (Case No. 35). His second case (No. 27) occurred in a nine-month-old infant and involved the sacrum, the tumor being of a lytic nature. Death occurred one year and three months after admission. A case of Ewing's tumor simulating osteomyelitis in a two-year-old infant was reported by Swenson and Teplick (1945). Extensive metastases involving almost every bone in the body, including marked decalcification of the spine, was noted within two months after admission. An incidental finding of interest in this case was marked arterial calcification in the upper and lower extremities. These authors speculated that the rapid bone destruction produced hypercalcemia with subsequent metastatic calcification of the vessel walls. On 2 occasions the serum calcium had been reported as being over 12 mg. per cent.

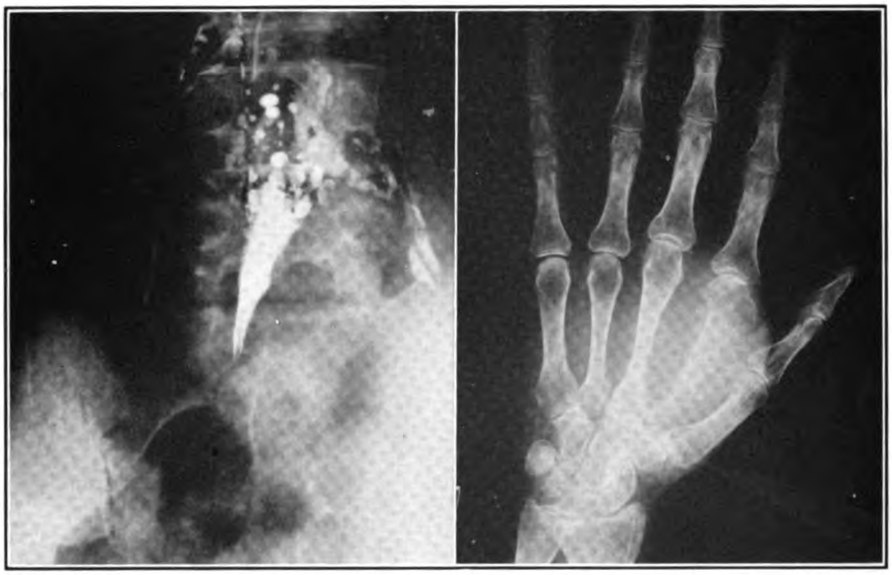

FIG. 219.—Ewing's tumor of the second metacarpal bone with metastases to the left side of the spinal canal at the lumbosacral interspace, displacing the pantopaque column to the right.

A series of 91 histologically proven cases of which the end result after five or more years of observation was known in 73 cases, was reported by Coley, Higginbotham and Bowden (1948). These authors found that only 3 out of the 73 cases survived five years or more regardless of treatment, whether *x*-ray or surgical or both. They mentioned that neurological complications were frequent and disturbing. In 14 cases studied at necropsy, vertebral metastases were encountered in 4, and in 74 cases vertebral metastases were suspected on clinical grounds in 21. In 3 cases pathologic fractures of the vertebræ were observed. In 3 cases the site of the primary lesion was in the sacrum, and none were noted primarily in the spine.

In our own material there were 2 verified cases of spinal Ewing's tumor. In the first, a ten-year-old boy, the eighth thoracic vertebra had a lytic lesion involving the left lateral aspect of the body. Adjacent thereto a large soft tissue swelling could be demonstrated. The neoplastic nature of the lesion was evident, but the ultimate diagnosis was made from material obtained at biopsy. The initial complaint was girdle pain at the level of the seventh rib, which had been present intermittently for about a year but had become rapidly progressive for a few days before admission to the hospital. Other neurologic signs included increased reflexes in the lower extremities, loss of bladder control and progressive weakness which progressed to a complete paraplegia at admission. At laminectomy it was noted that tumor tissue had extended into the left side of the spinal canal epidurally from the sixth to the eighth thoracic vertebra, deviating the cord towards the right.

The second case was a twenty-three-year-old woman who had pain in the back following a fall, with numbness in the left buttock and pain radiating to the left posterior thigh. This patient had had a swelling over the right second metatarsal bone for thirteen years. Myelography disclosed a displacement of the pantopaque column from right to left at the level of the fifth lumbar vertebra. The caudal sac assumed a sharp tapered appearance, which has been associated with epidural metastases. At laminectomy a tense reddish mass deflecting the cord from the fourth lumbar vertebra downward towards the right was noted. This mass was entirely extradural and was well encapsulated. Biopsy showed the structure of this tumor to be endothelial myeloma. *X*-ray examination of the hand showed a destructive lesion involving the shaft of the second metatarsal bone consistent with the diagnosis of a Ewing's tumor. This patient did quite well for about three years following operation with *x*-ray therapy both to the spine and to the hand, with regeneration of the bone of the hand.

Interestingly enough, the plain roentgenograms of the spine before myelography were normal and reinspection of the films after operation failed to disclose any evidence of bony destruction. It is postulated that the tumor spread to the epidural space by means of the vertebral plexus of veins from a site too small to be identified radiologically.

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Reticulum Cell Sarcoma of Bone.—These are malignant growths which usually occur in early life and may run a fairly long course. As encountered in bone, the lesion is much the same as reticulum cell sarcomas elsewhere in the body. According to Coley, Higgenbotham and Groesbeck (1950) the tumor as encountered at operation is characteristically soft, friable, glistening and fleshy with frequent areas of necrosis. Its consistency may vary from a soft mass to one of rubbery density. The lesion is primarily destructive in nature, and occasionally bony reparative changes may be visible. A characteristic feature of the growth is said to be an ill-defined boundary with extension of the tumor into the adjacent soft tissues. However, it is usually necessary to resort to biopsy for diagnosis.

The exact nature of these growths is important from the viewpoint of management of the patient. Reticulum cell sarcoma of bone is quite radiosensitive and may even be radiocurable.

A case of reticulum cell sarcoma of bone originating in the vertebral column was reported by Edwards (1940). Sherman and Snyder (1947) encountered 2 such cases in a series of 17 patients, 1 involving the fifth cervical and 1 the fifth lumbar segment. In both cases the lesions involved primarily the vertebral body and were primarily destructive in nature, with pathological fractures of the vertebral body involved.

Coley, Higgenbotham and Groesbeck (1950) reported 2 cases which originated in the lumbar vertebræ, and also mentioned that reticulum cell sarcomas originating elsewhere in the body may metastasize to the spine, as occurred in 2 of their other cases. In their report of a series of 17 verified cases, they mentioned 1 in which the tumor involving the body, pedicles, laminæ and transverse processes of the fifth lumbar vertebra recalcified after roentgen treatment. This patient was well three years afterward. In a review of 25 cases of primary reticulum cell sarcoma of bone, MacCormack, Ivins, Dahlen and Johnson (1952) found 1 case originating in the seventh thoracic vertebra.

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Spinal Lymphomas

It was formerly believed that lymphogranulomas spread only through the lymphatic system. At present, their spread through the reticulo-endothelial system is well recognized, and some believe that the disease should be considered one of the reticulo-endothelial system itself. The mechanism for skeletal involvement includes three possibilities: direct invasion, hematogenous spread, and transference of an agent capable of initiating a focus *in situ* (Falconer and Leonard, 1948).

According to autopsy reports the bones may be involved in 40 per cent of cases. The radiologic changes are much less conspicuous because the lesions are invisible unless sufficient destruction of cortical or medullary bone produces either a rarefied lesion or an actual collapse of the affected vertebral body. Coles and Schulz (1948) reported that of 288 cases of malignant lymphoma of all varieties, 13 per cent showed involvement of bone, the spine being the most common site. Mazet (1951) reported that of 28 cases of Hodgkin's disease examined post mortem 20 had lesions in the vertebræ. Of these only 1 revealed changes demonstrable radiologically. Of 8 cases of lymphosarcoma, 6 presented bone changes at necropsy, but none on radiologic examination of the bone. In 6 cases of reticulum cell sarcoma of the vertebræ, 4 showed obvious bone changes at necropsy, but only 1 had radiologically visible bone destruction. In a case of Hodgkin's disease reported by Húlten (1927) the involved vertebra presented a dense, white appearance designated as an ivory vertebra. Dresser and Spencer (1936) also mentioned that a dense vertebra may be seen in Hodgkin's disease.

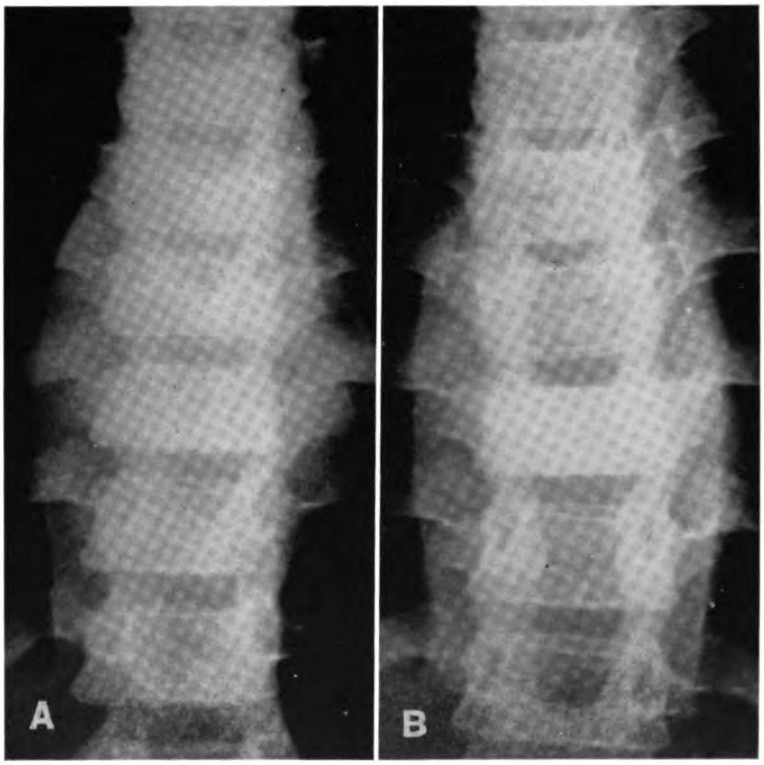

FIG. 220.—Lymphoblastoma involving the body of the eighth thoracic vertebra, with pronounced paravertebral soft tissue swelling. Five months later, following laminectomy for relief of compressive symptoms and *x*-ray therapy, the involved vertebra has increased in density. The paravertebral tumorous extension remains prominent. Exitus about three months thereafter.

In my experience, a dense vertebra was encountered in a child with Hodgkin's disease with spinal cord compression requiring laminectomy. The plain roentgenograms revealed paraspinal swelling bilaterally in addition to the dense vertebral body. However, this should not be regarded as characteristic of the disease.

Involvement of the central nervous system by lymphoblastomatous changes are quite well known. Verda (1944) reported that malignant lymphomas may arise in the epidural space itself, or migrate from nearby regional lymph nodes and present the same symptomatology as other cord tumors. Vietz and Hunter (1933) reported a case of lymphoblastoma causing paraplegia, requiring laminectomy of the third, fourth, fifth and sixth thoracic vertebræ. Beneath the fourth and fifth lamina an irregular granular mass was found extradurally compressing the cord. There was no involvement of bone. In this patient a rapid onset of paraplegia indicated

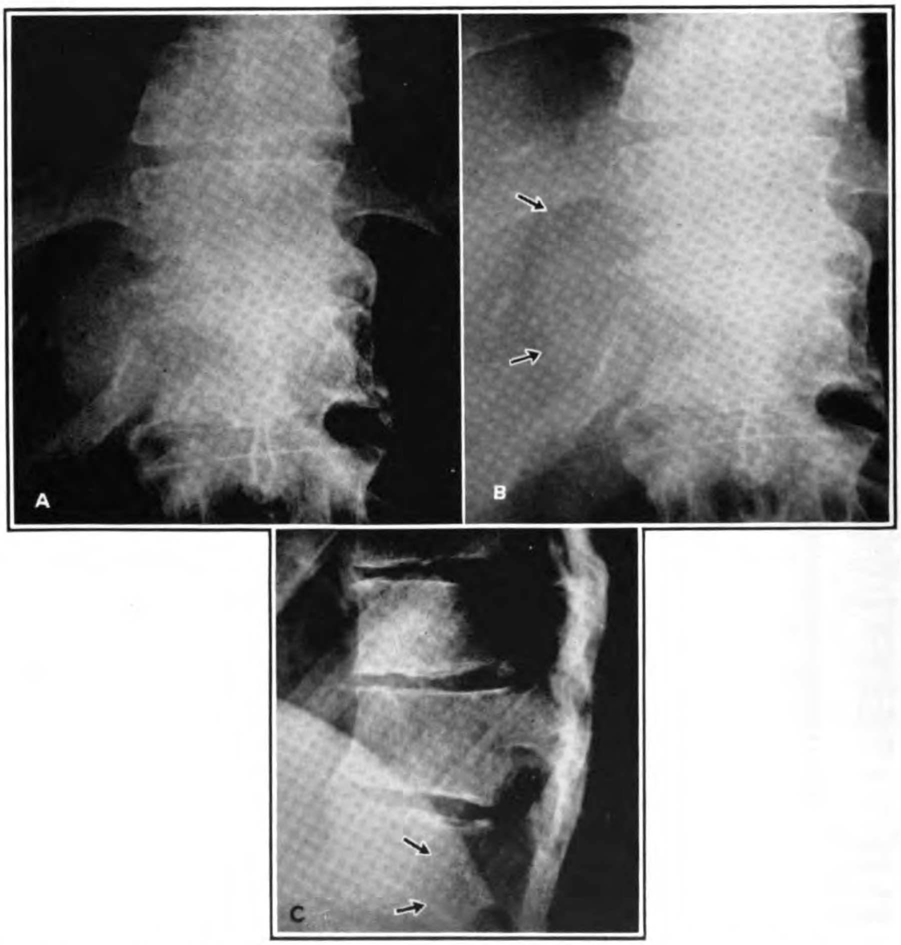

FIG. 221.—*A*, Hodgkin's disease of the tenth and eleventh thoracic vertebræ, with destruction of the right pedicle and the lateral margins of the vertebral bodies. *B*, Same case, soft tissue roentgenogram. Note the paravertebral soft tissue swelling (arrows). *C*, Same case, lateral roentgenogram. There is destruction of the posterior aspect of the body of the tenth thoracic vertebra (arrows).

the necessity for early operation. Compression of the spinal cord by Hodgkin's disease was encountered by Smith and Stenstrom (1948), who reported that of 305 cases of Hodgkin's disease 11 presented symptoms of spinal cord compression. Of these, 8 were verified at operation, and 3 were considered accurately diagnosed on clinical grounds.

Myelography in several of the cases of Smith and Stenstrom revealed obstruction, and laminectomy disclosed firm nodular extradural tumor masses. In their series radiologic examination of the spine revealed bone changes in 5 cases and myelography localized an area of block in 3 cases. The chief change seen on the plain films was erosion of the pedicles, but invasion of the vertebral bodies and destroyed laminæ were also present. Of the 8 cases with verified epidural masses, 4 had cervical, mediastinal or retroperitoneal nodes. In 2 cases the spread to the spinal canal was established at laminectomy to be by way of the intervertebral foramina. In 3 cases the dura itself was invaded.

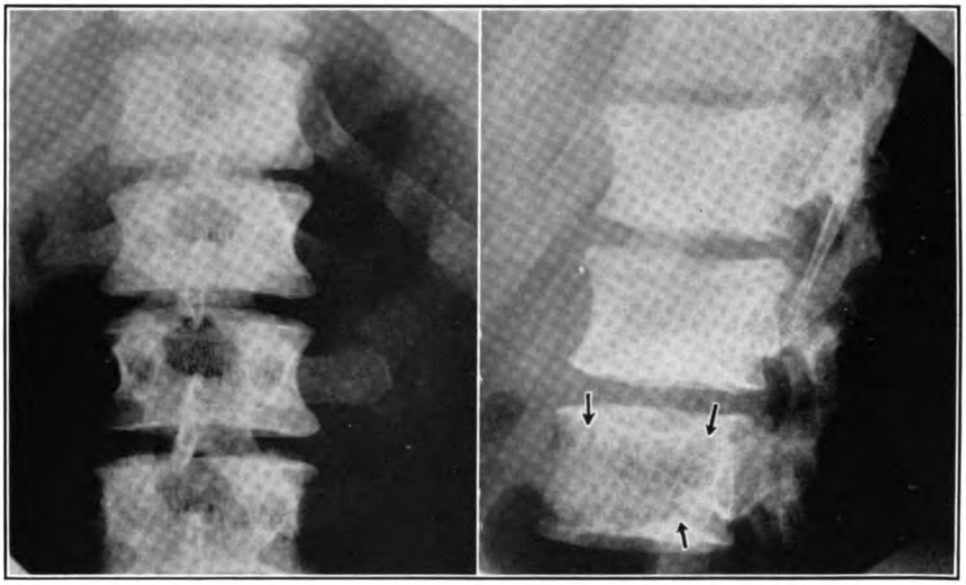

FIG. 222.—Hodgkin's disease of the body of the second lumbar vertebra. There is moderate loss of density and irregular medullary markings of the vertebral body, better seen on the lateral view (arrows). Note the loss of definition of the anterior aspect of the vertebra.

The radiologic aspects of spinal extension of lymphomatous tumors present a varied picture. In some cases the extension may simulate metastatic disease of any origin, with vertebral decalcification, loss of contour sharpness and varying degrees of collapse. In others the process may be purely lytic, the rarefied focus being sharply circumscribed from the rest of the bone. There may also be extension of lymphomatous tissue into the adjacent soft tissues. In still other instances there may be a mixed process, with small foci of increased bony density within the lytic areas, with relatively little change in the size or configuration of the vertebral body. Destruction of the pedicles and laminæ should be sought. In 1 case, we observed a reticular destruction of bone involving the lower cervical and upper dorsal segments (Fig. 232). This patient had signs of cord compression. He refused myelography. His symptoms were relieved after *x*-ray therapy.

It is frequently possible to identify the level of involvement in patients with Hodgkin's disease who have a combination of central nervous system disturbances with obvious bony changes. However, in the absence of visible bone destruction one should not hesitate to employ myelography whenever clinical symptoms indicate the possibility of central nervous system compression. While the classical picture encountered is that of a partial or complete block, we have also observed cases in which the deformities were semicircular, or presented an irregular curvilinear defect along the lateral aspect of the pantopaque column. This, of course, is dependent upon the structure of the intraspinal lymphomatous implant or extension.

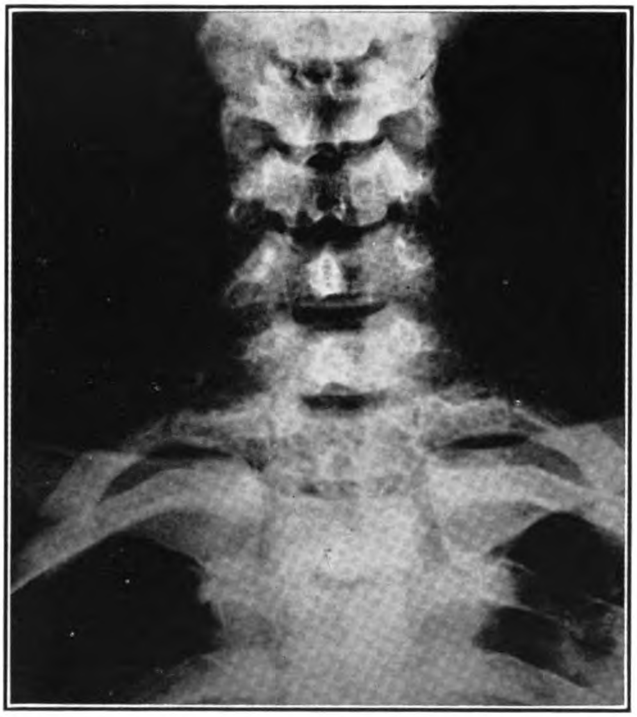

FIG. 223.—Hodgkin's disease of the cervical and the upper dorsal vertebræ. There is a web-like appearance of the involved bony structures.

In 1 of our cases the patient was considered to have a herniated disc, and myelography showed a fairly characteristic defect at the lumbosacral space. However, further investigation disclosed lymphomatous disease in this patient, and operation was delayed until a sufficient course of deep *x*-ray therapy could be administered. This patient had recession of symptoms following the *x*-ray therapy, so that operation was not required.

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SARCOMAS OF THE VERTEBRAL COLUMN

Osteogenic sarcoma arises from the primitive bone-forming mesenchyme. According to Lichtenstein (1952) the proliferating connective tissue in an osteogenic sarcoma is often quite anaplastic, and produces both tumor osteoid and bone tissue directly. Occasionally the proliferating connective tissue may develop cartilage which rapidly undergoes further differentiation into bone. Osteogenic sarcomas appear more often in males, and usually appear in adolescent and early adult life. However, cases may be encountered in adults, some of whom have Paget's disease. While the usual sites of origin are in the long bones, particularly in the vicinity of the knee and shoulder, such tumors also arise in the pelvis or in the spine. It is believed that osteogenic sarcomas first appear in the cancellous bone tissue, and gradually erode their way through the cortex of the bone, perforating the periosteum and expanding into the adjacent soft tissues. It is during this stage that the destruction of bone becomes visible roentgenologically and elevation of the periosteum may be demonstrated.

In some cases the bone forming tendencies of the tumor manifest themselves by calcifications in the depths of the tumor. However, as the tumor expands into the adjacent soft tissues foci of bone formation can be demonstrated in the periphery of the tumor as well as in its center. Extension of the tumor through articular cartilage is rare. Lichtenstein stated that the histologic pattern of osteogenic sarcoma is very variable, and based his diagnosis on the presence of a frankly sarcomatous stroma and the direct formation of tumor osteoid and bone by this malignant connective tissue.

In addition to the bone forming propensities of osteogenic sarcoma one occasionally encounters such a tumor which is predominantly lytic. In these cases the tumors are characterized by the coexistence of necrotic softening and hemorrhage with little or no evidence of tumor osteoid or pathologic bone formation. Roentgenologically these tumors may be extremely difficult to identify and the lytic sarcomatous nature of the disease may require histologic examination for its identification.

The occurrence of osteogenic sarcoma in the vertebral column is quite infrequent. Geschickter and Copeland (1949) identified 187 cases of sclerosing osteogenic sarcoma from a total of 750 osteogenic sarcomas. In this group the favorite site of origin was in the metaphyses of the long bones just adjacent to the epiphyseal lines. They mentioned that only a few isolated cases occurred in the ribs and vertebræ, the pelvis, and about the skeletal trunk. In reviewing their Table 17, 1 case of sclerosing osteogenic sarcoma of the sacrum is noted in the 109 cases listed. In Table 18, also headed as osteogenic sclerosing sarcoma, 1 case involving the second and third lumbar vertebræ was noted out of a total of 78 cases. Under a heading of osteogenic

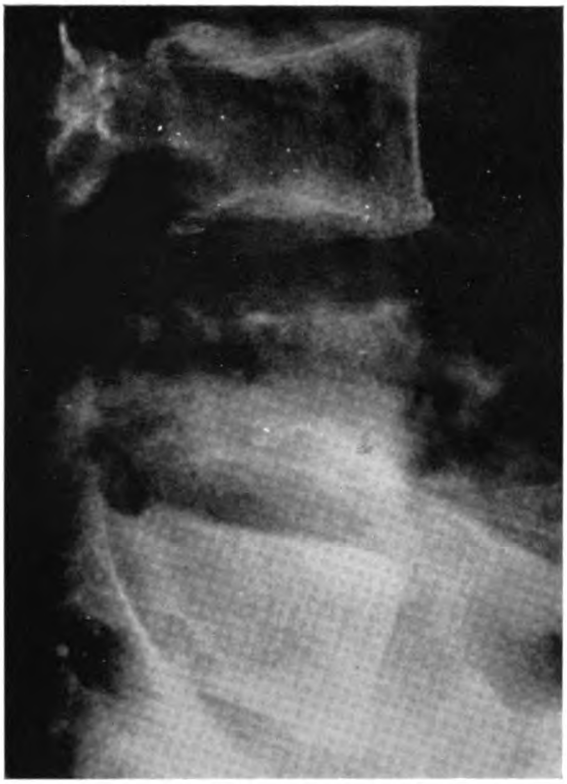

FIG. 224.—Osteogenic sarcoma of the body of the fourth lumbar vertebra, involving the superior and posterior aspects and extending into the pedicles.

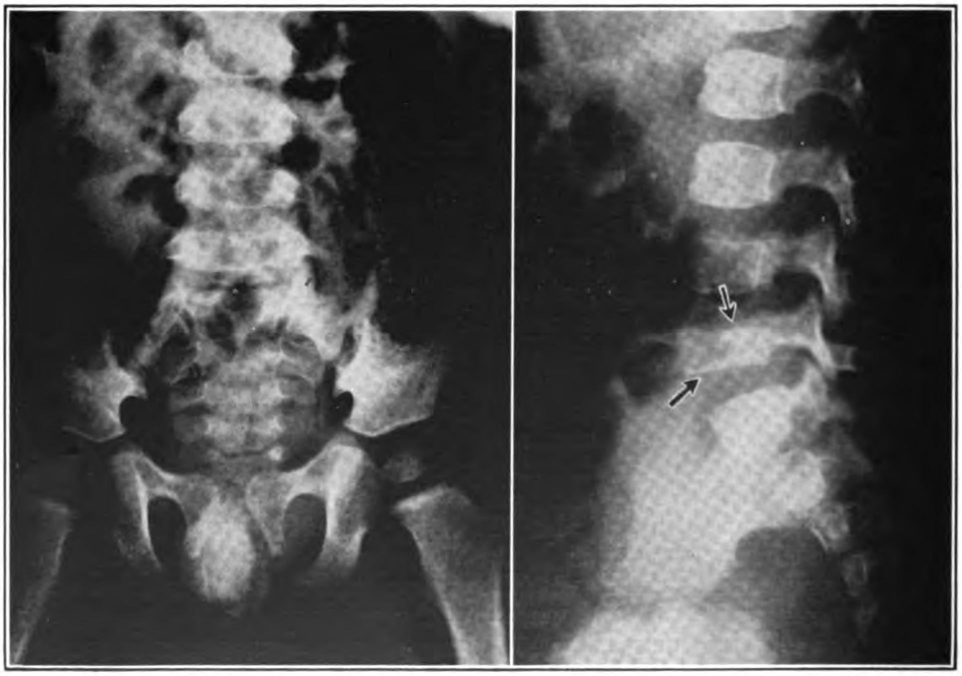

FIG. 225.—Sarcoma of body of the fifth lumbar vertebra in an infant, with flattening of the vertebral body (arrows). Slight bulging of the anterior border is present.

osteolytic sarcoma 1 case involving the vertebræ is listed out of a total of 62. Listed in Table 22, also headed as osteogenic osteolytic sarcoma, no cases of vertebral tumors were encountered in a total of 88 such cases.

In my experience 3 cases of osteogenic sarcoma of the osteolytic variety were observed. In 1 the fourth lumbar vertebra was involved, and the radiographic picture was that of a slightly flattened, decalcified fourth lumbar vertebra with slightly bulging edges. The intervertebral discs above and beneath this vertebra were normal and the appearance of the vertebra suggested a metastatic lytic neoplastic process. The true nature of the tumor was discovered at necropsy. The second was in an eighteen-month-old infant. The third case involved the fifth dorsal vertebra and the nature of the lesion was discovered following laminectomy because of spinal cord compression.

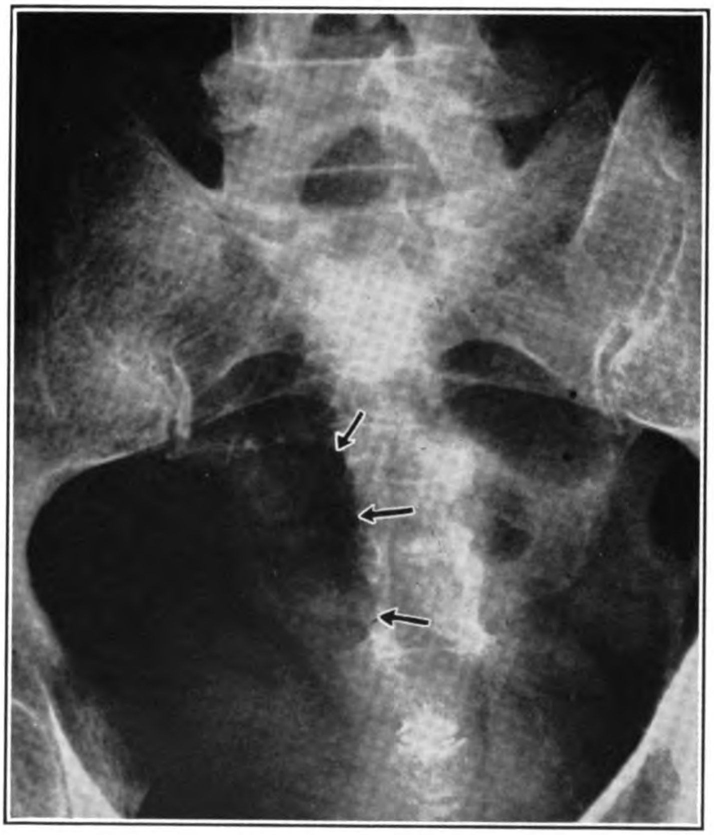

FIG. 226.—Invasion of the right side of the sacrum by a sarcoma originating in the adjacent tissues (arrows).

Fibrosarcoma.—Lichtenstein (1952) described fibrosarcoma of bone as a primary malignant fibroblastic tumor which does not form osteoid or bone, either locally or in its metastases. This tumor usually originates in cancellous bone, and manifests itself by outward growth causing perforation of the cortex and extension of the tumor into the adjacent soft tissues. Occasionally fibrosarcomas arise from the periosteum, and invade the bone by direct extension through the cortex. Stout (1948) considered these relatively benign growths of the fibroblasts, and presented an excellent illustration of a fibrosarcoma of the periosteum of the coccyx which pressed forward to indent the rectum. This tumor had not recurred sixteen years after resection.

Geschickter and Copeland stated that fibrosarcoma of bone, which they designated as fibrospindle cell sarcoma, may arise from the outer layers of the periosteum or the adjacent soft parts, and may invade the bones by extension from without. They did not regard this type of tumor as a primary osseous lesion. This view was debated by Lichtenstein, who preferred to

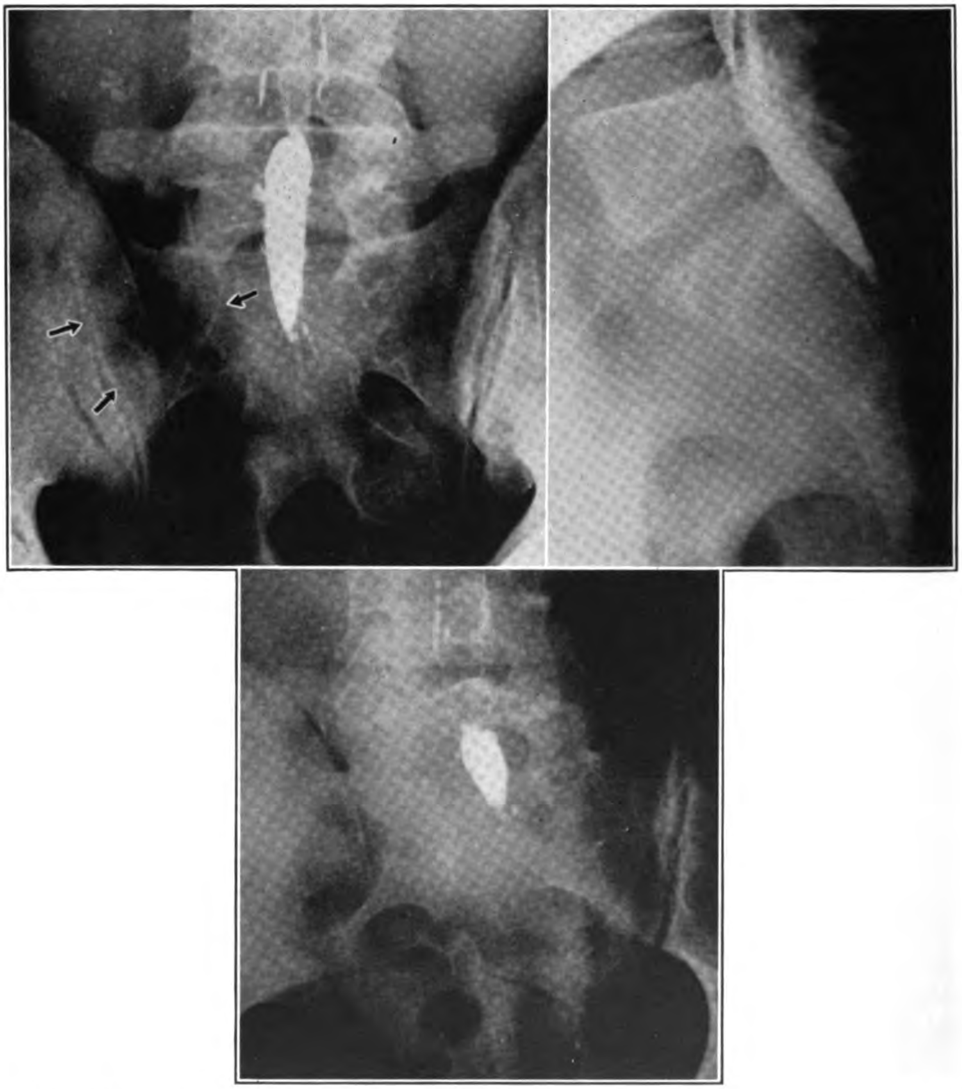

FIG. 227.—*A*, Fibrosarcoma originating in right superior aspect of the sacrum (arrows). Slight tapering of the caudal sac is present. *B*, Same case, lateral myelogram. No displacement of the pantopaque column is seen. *C*, Same case, thirteen weeks later. Note the increased contralateral displacement of the pantopaque remaining in the caudal sac. The destruction in the right upper sacrum is more marked.

regard osseous medullary fibrosarcomas as a widely variable group of tumors which may be highly malignant or relatively slow growing, depending upon their cellular characteristics.

In my series there were 5 cases which were described by the pathologists (the late Dr. Max Lederer and Dr. David Grayzel) as fibrosarcomas of bone. These occurred in young and middle-aged adults, and involved the fourth lumbar vertebra in 1 case, the fifth lumbar vertebra in another, and the upper portion of the sacrum in the remaining 3. From the radiologic viewpoint the lesion was characterized by a faint lytic process which under observation became progressively larger. In 3 cases myelographic examination was done because the clinical picture simulated that of a herniated disc. In these the distal end of the pantopaque-filled thecal sac was tapered and deviated contralaterally. At operation the change was found to be due to epidural extension of the tumor tissue from the bone malignancy.

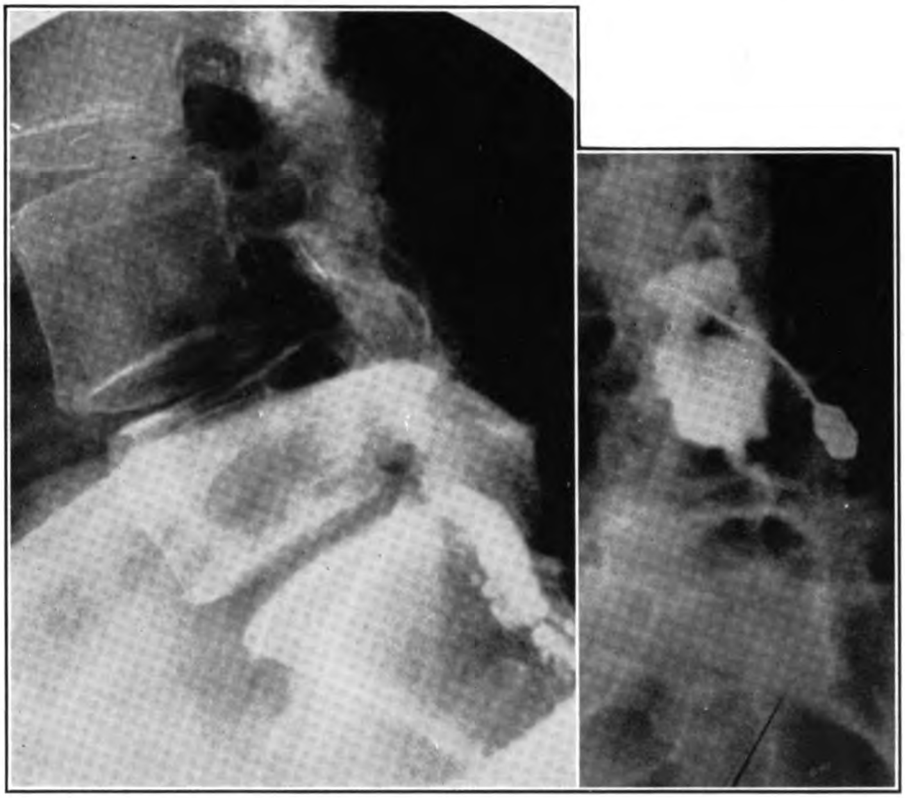

FIG. 228.—Fibrosarcoma of the fifth lumbar vertebra. There is destruction of the posterior aspect of the vertebral body. Complete block is noted on the myelograms at the fourth lumbar interspace.

It is worth mentioning that in 2 of these cases the preliminary roentgenograms made before myelography, had been considered within normal limits and the lesion was identified only after the operative findings were known.

Chondrosarcoma.—These are malignant tumors of mature cartilage which not infrequently appear as degenerative lesions in benign enchondromas, or less often, as malignant degeneration of the cartilaginous cap of an osteochondroma. Chondrosarcomas were classified by Lichtenstein and Jaffe (1943) as central, originating within bone, while those which appeared as malignant degeneration of the cartilaginous cap of an osteochondroma were referred to as peripheral chondrosarcomas. These tumors usually occurred in adults between thirty and fifty years of age. The most common site for this tumor was in the long bones, the pelvis and the ribs, but cases have been reported in the spine.

In our own experience we have observed 1 instance in which a chondroma arising from the lamina of the seventh dorsal vertebra underwent malignant degeneration and eroded into the body of that vertebral segment. The tumor also extended into the adjacent soft tissues. The presenting picture in this patient was of a slowly progressive nature, starting with pain in the back and eventually terminating with paraplegia. This patient, who was a twenty-seven-year-old male, was afflicted with hereditary multiple exostoses and undoubtedly one of these lesions was the original source for the development of the osteochondrosarcoma which caused his death. Another involving the vertebræ appeared in 1 of our patients with hereditary multiple exostoses. The development of chondrosarcoma on the lesions of Ollier's disease has also been described.

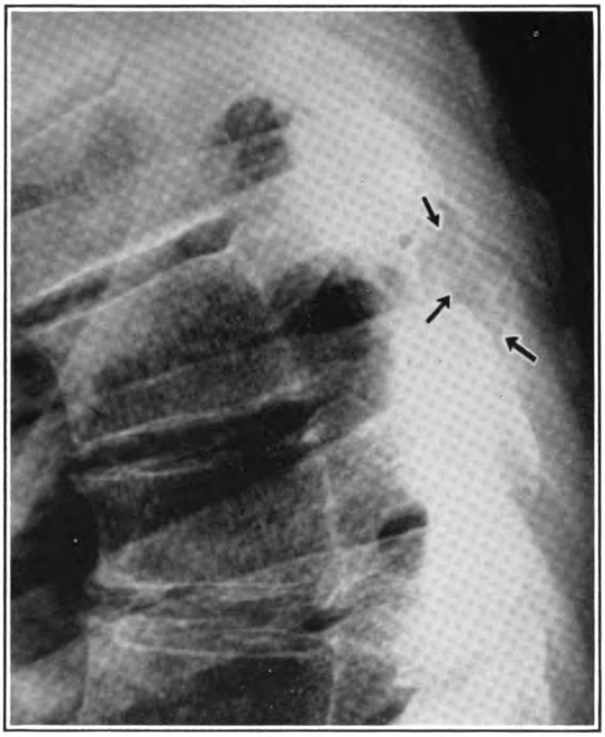

FIG. 229.—Osteochondrosarcoma originating in the base of the spinous process of the seventh thoracic vertebra (arrows).

Chondrosarcomas of bone are relatively slow growing tumors which metastasize late. They tend to be locally invasive for long periods of time before penetrating the bony confines and extending into the adjacent tissues or metastasizing distally. This occurred in another of our cases in which the sacrum and coccyx were affected.

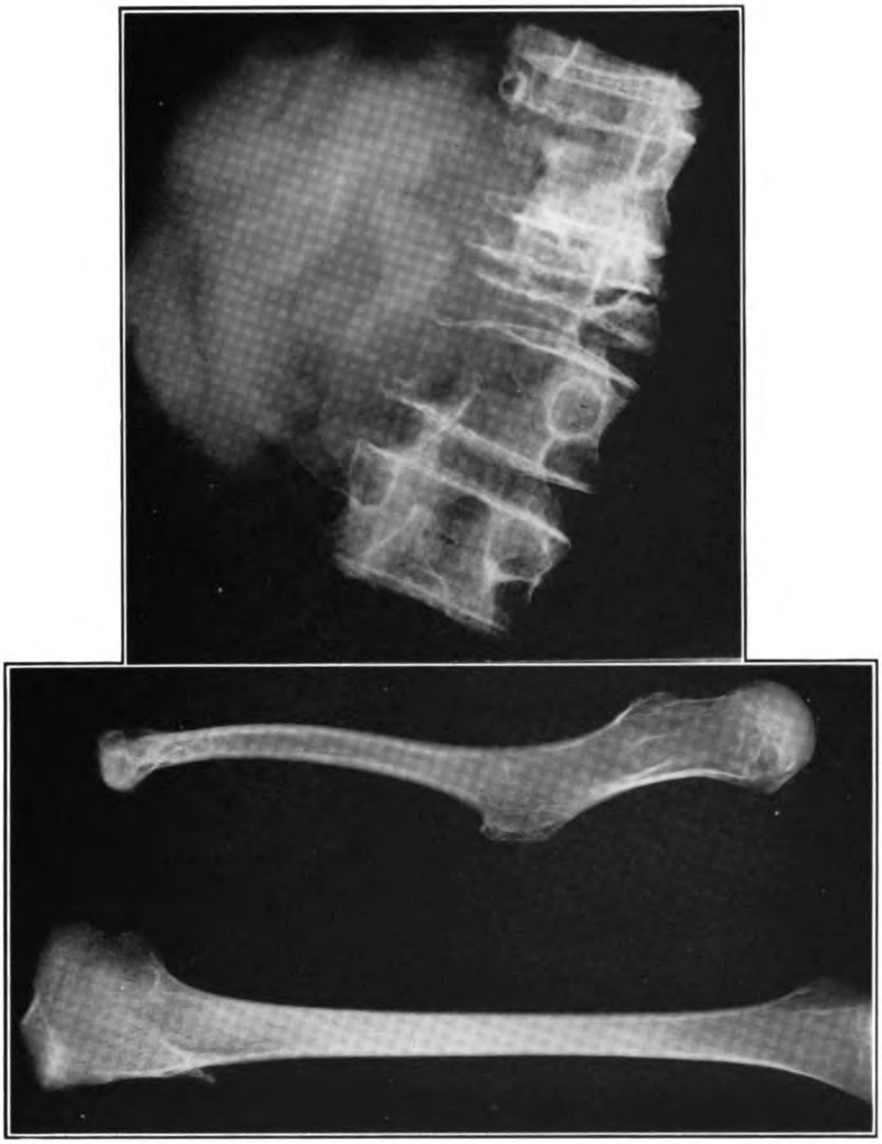

FIG. 230.—Roentgenogram of necropsy specimen of sarcoma destroying second, third and fourth lumbar vertebræ. Note the large paravertebral soft tissue neoplasm. The long bone specimens show multiple hereditary exostoses, and it was believed that the spine sarcoma originated from one of these even though no other exostoses could be found in the vertebræ.

An unusual case of a chondrosarcoma extending into the abdominal venous trunks from the lower part of the vertebral column was described by Ernst (1900). In a review of 40 cases of chondrosarcomas of bone O'Neil and Ackerman (1952) described 2 cases involving the spine, one arising from the spinous process and lamina of the second cervical vertebra, and the other from the body of the sixth dorsal vertebra.

Angiosarcoma.—These are malignant tumors of blood vessels which are better designated as hemangio-endotheliomas. The usual site of occurrence is at the proximal ends of the long bones, particularly the femurs and humeri, the hands, feet, pelvis, and rarely the vertebræ. The tumor is predominantly a destructive one which gives rise to a wide expansion of the bone, and presents a rather characteristic appearance which has been described as "soap bubbles."

The neoplasm is one which is formed by blood vessels, and is characterized by an interwoven network of anastomosing vascular channels. The increased circulation in the tumor may give rise to an audible bruit, and sometimes a pulsation within the tumor may be detected on physical examination.

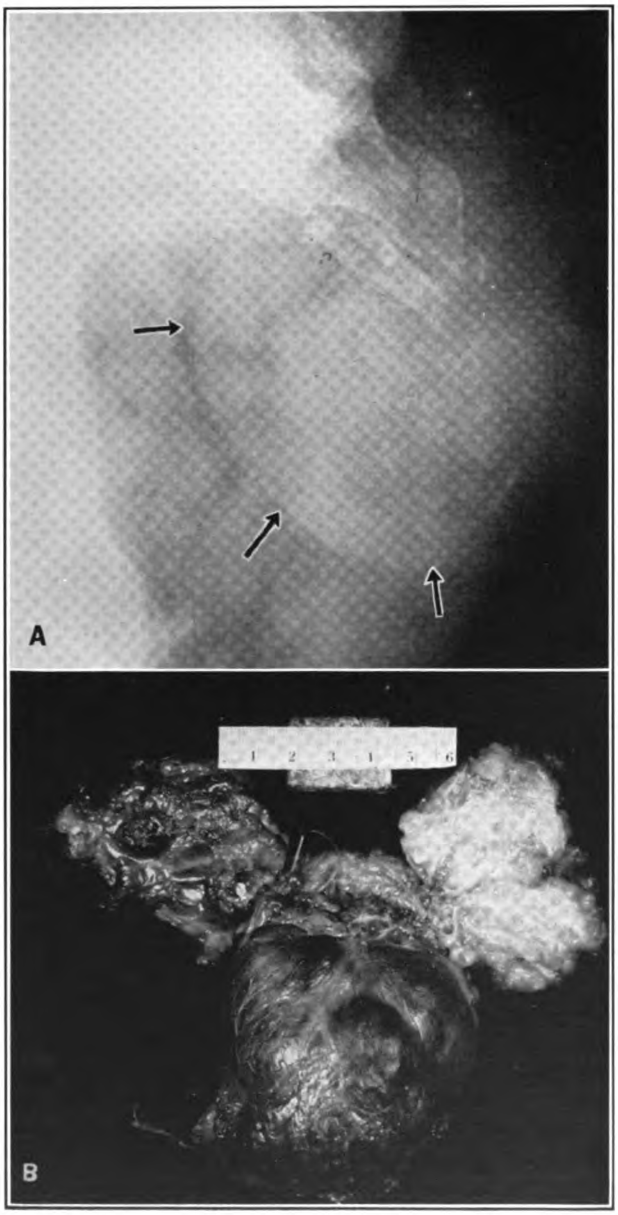

FIG. 231.—*A*, Osteochondrosarcoma of the sacrum with destruction of the distal half of this bone. A large soft tissue mass is seen (arrows). *B*, Same case, Operative specimen.

The incidence of hemangio-endotheliomas in the spine must be infrequent. I have been unable to find any specific reference to this tumor as involving the vertebræ except for mention by Brailsford who included the vertebræ among the sites of origin.

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CHORDOMAS

These relatively infrequent, slowly growing malignant tumors occur mostly at the cephalic and caudal ends of the vertebral column, and originate from remnants of the notochord. It has been shown by Horowitz (1941) in a study of 50 human embryos and fetuses, that the topographic distribution of heterotopic chordal vestiges correspond closely to the sites of occurrence of chordomas. Before him, Schmorl (1928) had demonstrated chordal remnants in 9 adult cadavers in dissection of over 3000 human vertebral columns. In 6 of these the chordal remnants were placed posteriorly, in 2 centrally, and in 1 instance laterally in the vertebral bodies. While these tumors occurred most frequently in the upper and lower ends of the spinal column, particularly in the region of the clivus Blumenbachii and the sacral-coccygeal region, they also were found in the cervical, thoracic and lumbar spine. They apparently never originate in the intervertebral discs.

In a study of 59 cases by Dahlin and MacCarty (1952) 15 chordomas were found in the vicinity of the clivus Blumenbachii, and 32 in the vicinity of the sacrum. Of the remainder, 9 occurred in the cervical region, 2 in the fourth lumbar vertebra and 1 in the eleventh thoracic vertebra.

Wood and Himadi (1950) in a report of 16 cases, encountered 7 in the clivus, 1 in the nasopharynx, 5 in the vertebræ, and 3 in the sacrococcygeal region. A review of the literature by Faust, Gilmore and Mudgett (1944) disclosed 252 cases. Of these 92 were cranial, including the spheno-occipital and nasopharyngeal regions, 34 were vertebral in location and 122 were caudal or sacrococcygeal in location. Four were mentioned as occurring in the other regions. Littman (1953) summarized the literature on sacrococcygeal chordoma and gathered 168 known cases in patients varying in age from infancy to over eighty years.

These growths are usually rounded or lobulated masses of soft gelatinous tissue containing areas of cystic degeneration, necrosis, hemorrhage and sometimes calcification. In their early stages the growths usually are well encapsulated, but as they grow penetration of the capsule may occur and the tumor tissue invades adjacent bony structures and extends into the soft tissues. This is particularly apparent in sacral chordomas, where large presacral and infra-sacrococcygeal soft tissue masses may appear. In the cervical areas, these tumors may project into the retropharyngeal region sufficiently to first manifest themselves by dysphagia or other symptoms of a retropharyngeal mass. Tumors occurring in the vertebral column in areas other than the proximal and distal ends usually arise within the vertebral body itself. Chordomas arising from the odontoid process of the second cervical vertebra have also been encountered. The illustrations of chordomas entering the posterior and anterior cranial fossæ published by Adson, Kernohan and Woltman (1935) are of considerable interest.

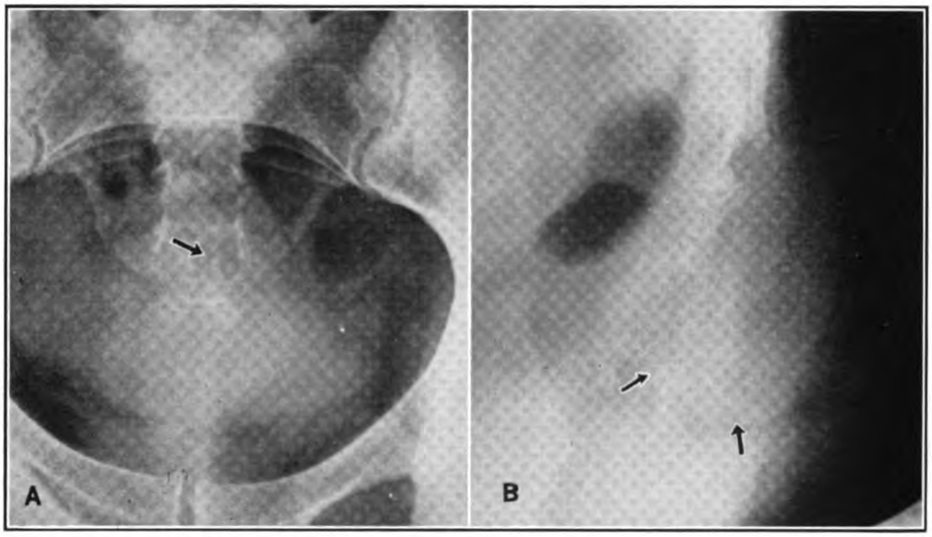

FIG. 232.—*A*, Sacrococcygeal chordoma, with destruction of coccyx. Note a central locule to the left of the midline in the lower sacrum (arrows). A soft tissue mass is seen just below the coccyx. *B*, Same case. Note the bony destruction and the adjacent soft tissue tumor (arrows).

Microscopically, chordomas present a picture simulating the embryonic notochord, being composed of large polyhedral cells with small nuclei and vacuolated cytoplasm distended with glycogen and mucin-like material. The cells are arranged in cords. The irregular clusters of cells are separated by a fine trabeculated stroma of connective tissue.

In an early stage, when the chordoma has not caused any destruction of its bony confines or protruded into any area in which altered function might bring the patient to the physician, the tumor may be symptomless. Once growth starts the clinical picture is dependent upon the area invaded as well as the extent of the tumor.

Sometimes chordomas reach a considerable size. In the case reported by Faust, Gilmore and Mudgett (1944) a sacrococcygeal chordoma extended into the lower quadrant of the abdomen. In our own experience 3 sacral chordomas reached a diameter of more than 15 cm.

Chordomas originating in the vertebral column may first manifest themselves because of pressure against the spinal cord or nerve roots. In those tumors originating low in the lumbar spine, a clinical picture highly suggestive of a protruded nucleus pulposus may be encountered, while in tumors originating higher in the vertebral column the tumors produce a clinical syndrome of spinal cord compression. Some chordomas simulate benign extradural growths or a destructive lesion of the vertebral body. If the vertebral body alone is involved, the roentgenograms show a rounded cavity, or there may be a mottled appearance. In the more advanced tumors there is a tendency towards collapse of the vertebral body. Richards and King (1940) reported a fifty-two-year-old male with a chordoma apparently arising in the third lumbar vertebra. Serial studies showed progressive destruction over a five-year period, starting with a decalcified vertebral body and discoid intervertebral discs above and beneath the lesion. The tumor advanced until there was complete collapse of the vertebral body, marked decalcification of the adjacent vertebræ and distortion of the interposed intervertebral discs. They noted that the amount of nerve involvement in this case was minimal for the size and location of the tumor. In the case reported by Robbins (1945), a forty-one-year-old man who died nine days after laparotomy, a large mass adherent to many abdominal organs was found. Microscopically the mass showed the two constant features of chordoma, an abundant mucous matrix and large physaliferous cells. Morris and Rabinovitch (1947) reported a chordoma in a twenty-seven-year-old woman with initial signs and symptoms of a protruded lower lumbar intervertebral disc. Myelography disclosed a block at this area. At operation an extradural mass was observed opposite the lumbosacral disc which could be followed into the body of the fifth lumbar vertebra.

Wood and Himadi (1950) described 5 cases of chordoma which involved the vertebral column. They noted that two or more adjacent vertebræ were involved in these cases, the tumor extending from the segment of origin to an adjoining vertebra by growth through the intervening intervertebral disc in each case. Bone destruction was a prominent feature, the final stages being compression fractures. In 2 cases osteoblastic reactions were observed, and expansion of a vertebral body extending into its transverse processes was seen in 1 case. In 2 cases myelographic examination showed complete obstruction. The pantopaque column tapered gradually to the point of obstruction in a manner usually seen with an epidural mass compressing the dural sac.

Poppen and King (1952) in a report on 6 cases of spinal chordoma, observed 3 in the lumbar or sacral regions. In all of these the tumors were extensive, and the spinal cord, the cauda equina and the nerve roots were compressed by extradural masses. At times the dura had been penetrated and intradural extension was marked. They believed that bone involvement occurred early in the course of the disease, and that the tumors spread widely in the intervertebral bodies and laminæ before they encroached on the cord. The tumor displayed an embryonal facility for easy penetration of the bone, the dura and nervous tissue. Of their 6 cases, 3 of which occurred in the cervical region, the diagnosis was not made preoperatively. All were thought to have metastatic malignancy. Gentil and Coley (1948) mentioned that in an analysis of reported case showed a correct *x*-ray diagnosis had been made in but 10 per cent of cases.

Chordomas in the thoracic region are rare. An interesting case was described by Hansson (1941) in a forty-five-year-old man with a level of hyperaesthesia at the level of the sixth and seventh dorsal segments. *X*-ray examination showed destruction of the neural arches, and operation revealed that the cartilage between the seventh and eighth thoracic vertebræ had been transformed into a bluish tumor mass which penetrated into the vertebral body. Bahlin and MacCarty (1952) mentioned that in their case of chordoma of the eleventh thoracic vertebra, decompression and subtotal removal were carried out. This resulted in a six-year remission, and reoperation for recurrence was necessary six-and-a-half years after the original operation.

We observed 1 case of chordoma worth recording. This occurred in a fifty-two-year-old woman who was seen because of pain in the low back extending into the right lower extremity. The clinical picture was compatible with a herniated disc. Myelographic examination was performed after plain roentgenograms showed no evidence of bony change. The myelogram showed a defect at the interspace between the fourth and fifth lumbar vertebræ. At operation a tumor was noted epidurally. This was a soft, gelatinous, friable mass which involved the body of the third lumbar vertebra inferiorly and the fourth lumbar vertebra superiorly as well as the intervertebral disc. The tumor was removed as completely as possible. Histologic examination revealed changes characteristic of chordoma. *X*-ray therapy was given postoperatively, and the patient was relieved of symptoms. She returned about one year later complaining of pain in the back. Further radiographic examination then disclosed a circular rarefied area within the body of the fourth lumbar vertebra without any distortion of the contours

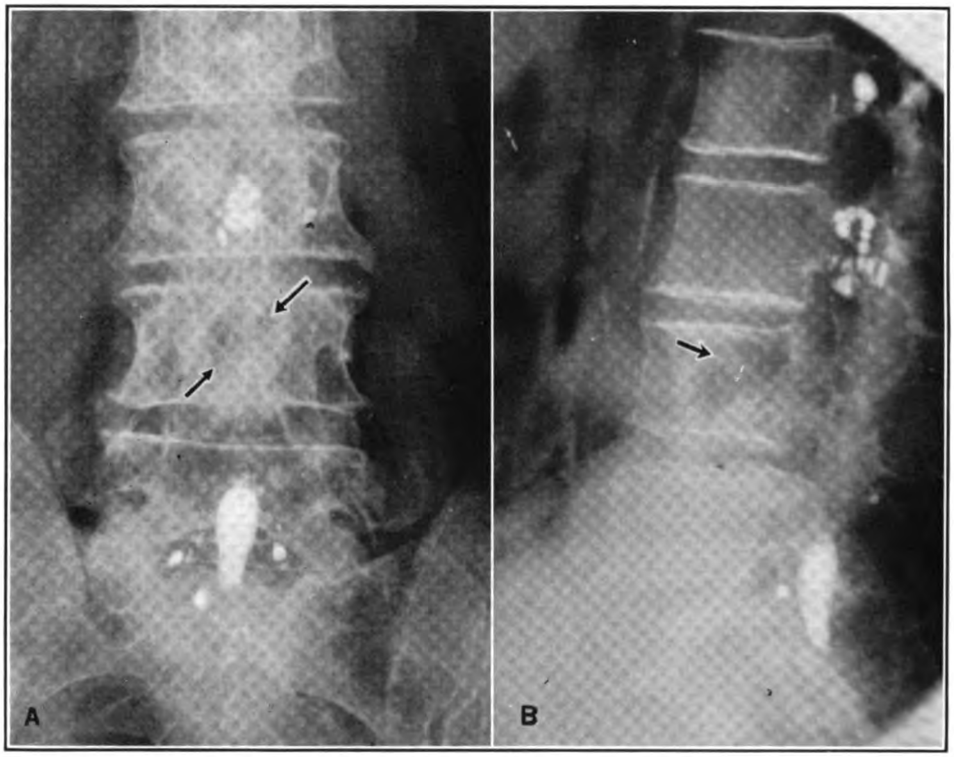

FIG. 233.—*A*, Chordoma originating in the fourth lumbar vertebra. A central area of rarefaction is seen (arrows). *B*, Same case. The rarefaction within the vertebral body is better visualized in this projection (arrows).

of this segment. *X*-ray therapy was again administered and the patient had symptomatic improvement.

While chordomas are known to be locally invasive, systemic metastases are relatively uncommon. In 1921 Lewis described 4 cases of chordoma, in 3 of which there were extensive metastases. Stewart (1922) commented upon the locally invasive and destructive properties of chordomas and demonstrated invasion of the lumens of the blood vessels. Willis (1930) reported a case of sacral chordoma with widespread metastases. Mabrey (1935) observed that metastases from chordomas occurred only in those tumors which arose in the sacrococcygeal region. He reported 10 cases with metastases from a group of 16 sacrococcygeal chordomas with post-mortem examinations. The regional lymph nodes were involved most often but blood borne metastases to the liver, lungs, pleura, peritoneum and other organs were described. Littman (1953) mentioned that metastases occurred in about 10 per cent of the cases of sacro-coccygeal chordoma.

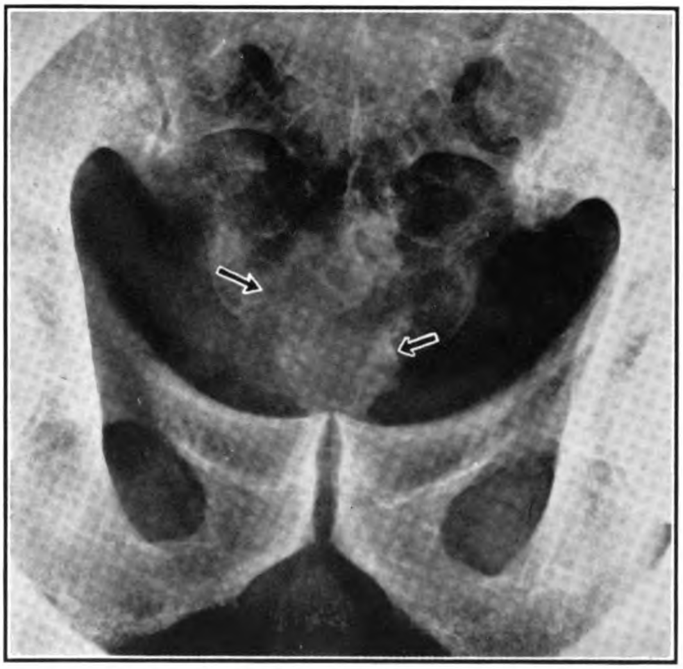

FIG. 234.—Sacrococcygeal chordoma, with irregular bands and islands of bone within the area of the tumor in the lower sacrum (arrows).

The roentgenologic changes associated with chordomas are predominantly of a lytic nature. Those tumors originating in the upper cervical region may destroy the bodies and occasionally the neural arches, producing changes in alignment of the cervico-occipital area. A fairly common site of origin in this rare condition is the odontoid process. Soft tissue swelling anteriorly into the nasopharynx may be an early manifestation of such a growth, and appears even when the destructive bone changes are minimal or even invisible. The intervertebral discs are well preserved. In a case mentioned by Sennett (1953) the cervical esophagus was displaced anteriorly and to the right by a soft tissue mass emanating from a cervical chordoma. Myelographically, varying degrees of block are noted.

Vertebral chordomas occurring in the thoracic or lumbar vertebræ usually produce bone destruction which cannot be differentiated accurately from other forms of destruction of a neoplastic nature. These growths are slowly progressive, and a tendency for dislocation or collapse of the vertebral body exists. Osteoblastic reactions are infrequent (Baker and Coley, 1953). Wood and Himadi (1950) mentioned that such changes were observed in 2 out of 5 cases of vertebral chordomas in their series. As noted in 1 of our cases, the plain films may be entirely normal, and still vertebral and discal involvement may be identified at operation.

Sacrococcygeal chordomas are most frequent, and present a varied roentgenographic appearance. Expansion of the sacrum as observed in the lateral view is a common feature in the early stages of this growth. By and large, the sacrum is slowly destroyed by the chordoma, and extension into the soft tissues as a relatively well defined mass can be noted on appropriate roentgenograms. The destroyed edge of bone from which the tumor emerges may lead to a correct interpretation in some cases. Remnants of calcification within the soft tissue mass may be present. Compression of the rectum and bladder by the soft tissue components of sacrococcygeal chordomas may produce symptoms, but the growths do not as a rule ulcerate through these structures.

From a diagnostic viewpoint much depends on the location and extent of the chordoma as well as its potential for entering the spinal canal. When considerable bone destruction is present the appearance often cannot be distinguished from that due to metastatic neoplastic disease. If paravertebral swelling is seen it mimics that seen with osteomyelitis of infectious or granulomatous origin. Tumors low in the lumbar canal simulate herniated discs, and when little bone destruction exists the diagnosis is made fairly often on histologic examination of tumor tissue removed from the epidural space. Similar tumors in the nasopharynx simulate primary tumors of this area such as lymphoepitheliomas or carcinomas, more so if there is destruction of the sphenoid. High cervical tumors, with or without intraspinal protrusion, likewise must be considered. Extension of chordoma into the pelvis and abdomen may simulate ovarian cysts or other tumors, and with spread a picture like that of peritoneal carcinosis of other origin appears. Tumors still confined within a capsule cannot be distinguished clinically from other presacral newgrowths. With destruction of the sacrum metastatic or primary newgrowths, presacral neurofibromas or meningoceles must be considered. The infrequent chordomas originating within the bodies of vertebræ may be confused with primary tumors such as osteoclastomas or osteoblastomas, and diagnosis is best established by biopsy.

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PRESACRAL TUMORS

In a discussion of the roentgenologic diagnosis of tumors involving the sacrum, Camp and Good (1938) divided these growths into three groups. Those arising within the sacral canal, of which the ependymal cell glioma is the commonest, cause a smooth loculated erosion of bone. The other two are tumors arising within the sacrum proper, of which chordomas and teratomas are the most frequent. Occasional cases of Ewing's tumor and various forms of sarcoma are seen.

Brindley (1945) observed that neoplasms in this location were of many pathologic types, usually arising from vestigial embryonic tissue. He commented that the potentialities of the sacrococcygeal region were based on the fact that the primitive node occurred there, and from it all fundamental systems of the body grow forward. During fetal life this remains for a long time a point of fusion of the central nervous system, the skeletal axis, segmental musculature and the postnatal gut.

The tumors encountered most frequently in addition to those mentioned above are giant cell tumors, neurofibromas and fibrosarcomas as well as metastases from primary lesions elsewhere. Anterior sacral meningoceles were discussed in a previous chapter.

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GIANT CELL TUMOR (Osteoclastoma)

In the past the term giant cell tumor has been used rather loosely, and there is still uncertainty as to what should be regarded as a giant cell tumor of bone. In Lichtenstein's opinion the growth is a distinctive neoplasm which apparently arises from non-bone-forming supporting connective tissue of the marrow which can readily be identified on the basis of cytologic details. The pattern presented by these tumors consists of a vascularized network of ovoid or spindle-shaped cells interspersed with multinuclear giant cells. As a result of his investigations, he separated these tumors from many others which have been confused with them. Chief among these are the xanthic, spindle cell, or healing variety, of giant cell tumors which are now designated as non-osteogenic fibromas of bone. The calcifying or chondromatous giant cell tumor is now designated as benign chondroblastoma of bone, probably representing an independent benign tumor of cartilage forming connective tissue, unrelated histogenetically to giant cell tumor. Another rather frequent entity which has been termed atypical subperiosteal giant cell tumor has been clearly identified as aneurysmal bone cyst. It is Lichtenstein's opinion that giant cell tumors of the spine are exceedingly rare, and that some of the lesions so designated in the past represent aneurysmal bone cysts.

The usual locations of giant cell tumors are in the distal end of the femur, the proximal end of the tibia and the distal end of the radius. Occasionally, tumors of this sort have been observed in the upper end of the humerus, the upper portion of the femur and of the fibula, and in the patella. These tumors rarely appear in individuals under twenty years of age, and are characterized by a lytic process with little or no bone production adjacent thereto. The cortex of the bone on the side of the tumor may practically disappear. At one time it was believed that the giant cell tumor characteristically presented a latticework of bony traversing strands across the lytic area, but this view has been abandoned. The difficulty of making a diagnosis of the histologic type of a tumor on the basis of the radiographic changes alone is apparent.

The question as to biopsy verification of histologic characteristics has largely been resolved in favor of such examination of any accessible lesion. Such a view was presented by Buschke and Cantril (1949). However they do not consider the vertebræ in this group. There are others, notably Brailsford (1944), who believe that the characteristic picture of giant cell tumor may be easily recognized on the roentgenogram. In his opinion excellent results can be obtained by *x*-ray therapy alone, and biopsy may possibly be hazardous in that stability of bone may be weakened, and there may be possible dissemination of tumor cells. He suggested that there was evidence that surgical trauma may incite malignant metastases in some cases. By the same token, there are others (Murphy, 1935; Gershon-Cohen, 1943; Cade, 1949; Prossor, 1949) who hold that *x*-ray therapy alone may be very effective, but Lichtenstein pointed out that to his personal knowledge several cases of sarcoma developed within an irradiated and ostensibly cured giant cell tumor after a long latent interval. Cahan, Woodward, Higgenbotham, Stewart and Coley (1948) have reported several cases of sarcomatous degeneration in irradiated bone as well.

While most giant cell tumors are benign, there is evidence to indicate that some are malignant even at their onset or that they eventually become malignant, particularly following some form of inadequate therapy. It is not possible to distinguish radiologically the so-called malignant forms of giant cell tumor from those which might be relatively benign. This constitutes another factor influencing the trend towards biopsy investigation before instituting therapy.

Coley (1935) reported a group of cases diagnosed as benign giant cell tumor from biopsy tissue and *x*-ray films which later underwent malignant change. He estimated that this malignant change might be expected in as high as 15 per cent of cases, a figure which appears to be somewhat higher than reported by other observers.

It is possible that a pathologic fracture alone may result in healing of such a lesion. A case of a giant cell tumor of the lumbar vertebra was reported by Murphy (1935) in which operation was deemed inadvisable and *x*-ray therapy was refused by the patient. Healing followed a pathologic fracture which presumably stimulated healing.

"Giant cell tumors" of the spine affect the vertebral bodies or the neural arches, alone or in combination. As a rule there is decalcification of the involved area, the lesion being demarcated from the normal bone by smooth edges which merge with the adjacent normal structures. The lesion may produce expansion of the involved vertebra with thinning of the cortex, and a variable extent of linear traversing bands may be present. These, however, cannot be considered as of definite diagnostic importance. If the giant cell tumor expands towards the spinal canal it may cause spinal cord or cauda equina compression.

In view of the changing concepts of the pathology of tumors of the vertebræ, it becomes quite difficult to accept without reservation reports of such growths in the literature. However, mention may be made of several should the reader wish to investigate further. Cases were reported by Murphy (1935), Jenkinson, Hunter and Roberts (1938), Gershon-Cohen (1933), Brock and Bogart (1945), Prossor (1949) and many others. Two cases with biopsy confirmation were recently reported by Hamsa and Campbell (1953).

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BENIGN CHONDROBLASTOMAS. (Epiphyseal chondromatous giant cell tumor of Codman).

While sometimes confused with osteoclastomas, these tumors are not related to them histogenetically. Benign chondroblastomas are classified among the benign tumors of cartilage or cartilage forming connective tissue, occur mainly in young males under twenty years of age, and tend to involve the articular ends of long bones, especially the humerus, femur and tibia, and crosses the epiphyses. The tumor is lytic, has poorly demarcated margins and may bulge byond the cortex without actually destroying it. Stippled calcific densities may be present within the tumor. It should not be confused with chondrosarcoma or osteogenic sarcoma. Chondroblastomas are curable, and do not undergo malignant degeneration. Its occurrence in the spine is rare, one such case being reported recently by Buraczewski, Lysakowska and Rudowski.

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ANEURYSMAL BONE CYSTS

As mentioned above, many bony lesions of the spine have been reported variously in the literature as primary giant cell tumors, giant cell variants, subperiosteal giant cell tumor, giant cell variants of bone cysts, osteitis fibrosa cystica, pulsating benign giant cell tumor, angioma, ossifying hematoma and even osteogenic sarcoma. Among these undoubtedly are many

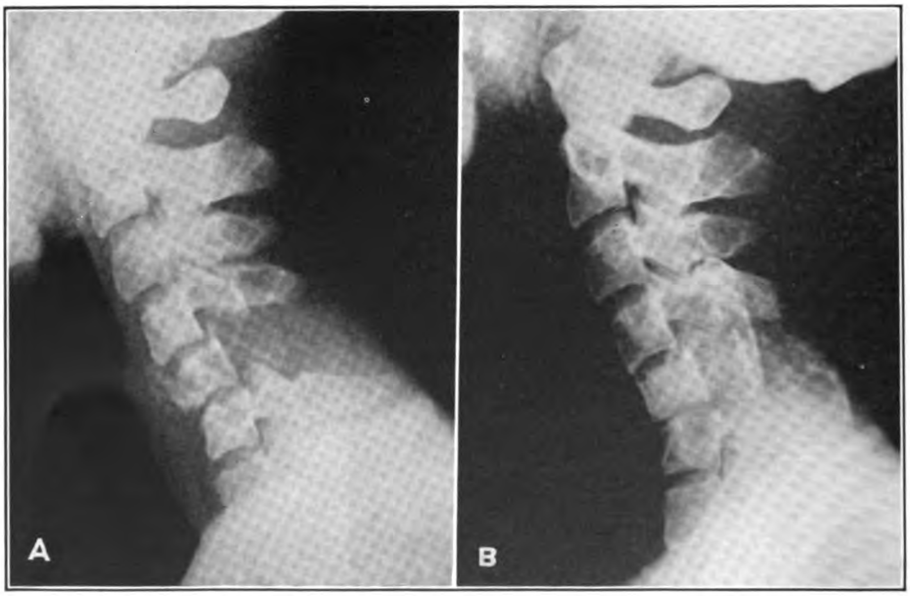

FIG. 235.—*A*, Destruction of the neural arch and spinous process of the fifth cervical vertebra. The vertebral body is also affected. This was believed to be a giant-cell tumor or aneurysmal bone cyst. *B*, Same case, four months after *x*-ray therapy. Extensive recalcification of the fifth cervical vertebra has taken place, with complete relief of symptoms.

examples of aneurysmal bone cysts (Besse, Dahlin, Bruwer, Svien and Ghormley, 1953). This condition was first brought into prominence by Jaffe and Lichtenstein (1942).

Aneurysmal bone cysts occur in children and young adults as solitary, localized expanded fibrous tissue lesions honeycombed with an enormously dilated vascular bed. These may appear in the long bones, in the short bones of the feet, the flat bones, the sacroiliac area, the calvarium and the

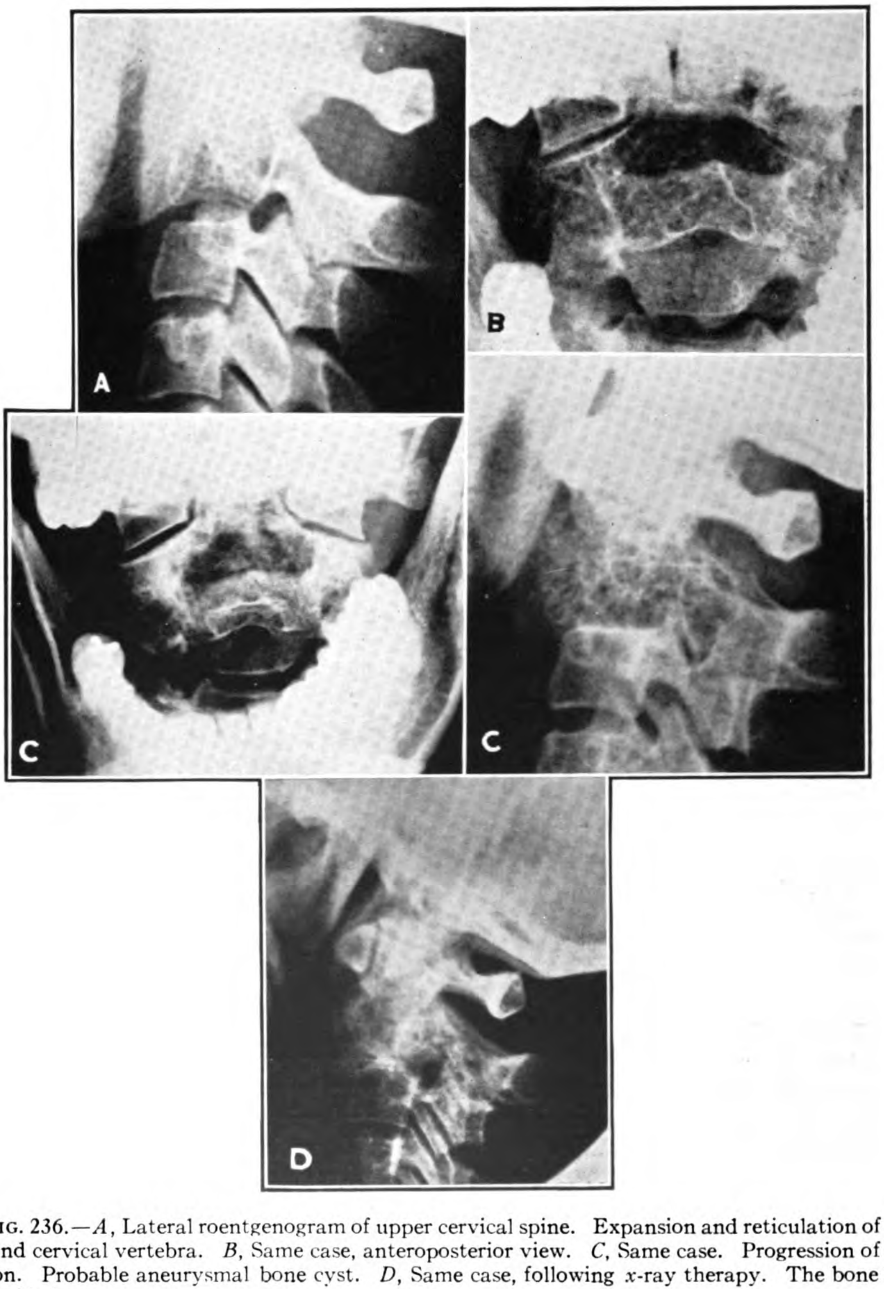

FIG. 236.—*A*, Lateral roentgenogram of upper cervical spine. Expansion and reticulation of the second cervical vertebra. *B*, Same case, anteroposterior view. *C*, Same case. Progression of the lesion. Probable aneurysmal bone cyst. *D*, Same case, following *x*-ray therapy. The bone has recalcified.

spine (Jaffe, 1950; Lichtenstein, 1950). Of 17 mentioned by Lichtenstein (1953), 4 were encountered in the spine. The neural arches of one or perhaps two adjacent vertebræ may be involved, although occasionally the body may be cystically transformed with a thin cortex. Trabeculæ may be visible across the thinned bone, but these probably represent endosteal ridges and spurs rather than true septæ. The normal cancellous bone is replaced by fibrovascular tissue with numerous large dilated vascular pools and channels. Encroachment on the spinal canal may result in spinal cord compression.

The radiographic appearance of vertebral aneurysmal bone cysts is one characterized by localized rarefaction and bulging of the involved bone, with thinned cortex and sometimes small focal deposits of new bone and calcifying osteoid tissue.

Aneurysmal bone cysts are amenable to surgical treatment or *x*-ray therapy. In 2 cases seen by us, and at first believed to be giant cell tumors, healing occurred after deep *x*-ray therapy. These patients were followed from five to twelve years. In 1 case, in which the spinous process and neural arch of the fourth cervical vertebra was involved, and which was not confirmed by biopsy, healing occurred within three months after the course of *x*-ray treatment had been concluded. At first a web-like stroma outlined the restored contours, and ultimately dense calcification occurred in this nineteen-year-old male patient. The second patient was a fourteen-year-old girl in which the second cervical vertebra was markedly decalcified, bulging, and traversed by thin septæ. *X*-ray therapy was followed by restoration of the bone, which remained relatively ballooned but reassumed a good osseous density.

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Hemangiomas

While hemangiomas of the vertebral column are among the more frequent tumors affecting this region, they are, nevertheless, relatively infrequent. From the viewpoint of incidence in pathologic specimens, Schmorl reported that of 3829 spinal columns examined, an incidence of 10.7 per cent of angiomas was noted. This series consisted of 1948 males and 1881 females, and the angiomas were more frequent in females. In 66.5 per cent of cases, only 1 angioma was demonstrated. In the remaining 32.8 per cent, 2 to 5 angiomas were seen, and in 3 cases even more were observed. In all, 579 different angiomas were encountered. Of these 32 occurred in the cervical region, 350 in the thoracic region, 170 in the lumbar region and 27 in the sacrum. The most frequent sites were the twelfth thoracic vertebra (47 cases), the fourth lumbar vertebra (38 cases), the first lumbar vertebra (37 cases), the second lumbar vertebra (36 cases), and the third lumbar vertebra (36 cases). Most of these were incidental findings in routine necropsy examinations, and were without clinical symptoms. They varied in size from a pinhead to lesions involving almost the entire vertebra, extending to the neural arches, or any portion of a vertebral segment.

Töpfer (1928) reported that hemangiomas of the vertebral bodies were encountered in approximately 12 per cent of 200 patients examined at necropsy. However, Lichtenstein (1952) made the point that occasionally an incidental finding at autopsy was a circumscribed, roundish, bright red hemangioma-like focus within the spongiosa of a vertebral body. It was more likely that these represented venous localized spaces rather than true tumors. He also indicated that lesions considered as angiomatous may actually be aneurysmal bone cysts.

Angiomas are dark red in color, and if extensive enough, may cause slight expansion of the entire vertebral body, with bulging of angiomatous tissue into the epidural space. Angiomatous tissue destroys the trabeculæ of the vertebral body, so that the remaining trabecular structures become thickened, producing the vertically parallel appearance of the remaining cancellous strands noted radiologically.

Ghormley and Adson (1941) divided angiomas into four groups including those which were asymptomatic, those which presented local signs and symptoms without spinal cord compression, those which presented symptoms of spinal cord compression without paraplegia, and those associated with paraplegia. The coexistence of compression myelopathy with vertebral hemangioma has been known for a long time. Soon after the original description of the radiologic appearance of hemangiomas by Perman (1927), Bailey and Bucy (1929) described a case of cavernous hemangioma of a vertebra associated with compression myelopathy. By 1942 Ferber and Lampe were able to collect 52 cases of vertebral hemangioma with spinal cord compression. In a discussion of the possible causes of compression of the spinal cord, Schlezinger and Ungar (1939) pointed out that this condition could be brought about by a ballooning of the vertebra caused by thrombosis and edema associated with osteoporosis and subsequent formation of bone by the extension of angiomatous tissue to the epidural space. This occurred in 8 of the 40 cases reviewed by them. Fracture of the involved vertebra causing spinal cord compression was encountered in only 4 cases in the literature, indicating the rarity of this complication.

An interesting point was made by Karshner, Rand and Reeves (1939) who described an epidural hemangioma associated with a hemangioma of a dorsal vertebra giving rise to compression myelitis. In this patient there were hemangiomatous nevi in the same dorsal segment as the epidural hemangiomas. Brøbeck (1950) reported a sixteen-year-old male patient with a spastic paraparesis of the lower limbs due to a hemangioma of the seventh dorsal vertebra. Operation disclosed extradural vascular tumor which involved the neural arch, and compressed the spinal cord from its dorsal aspect. This patient had complained of prolonged insidious development of disturbance in gait, increasing disturbances in sensibility and severe pain. Brøbeck mentioned that pain was a rare and often a late symptom and may sometimes occur in periodic attacks, presumably brought about by congestion in the cavernous blood spaces of the tumor. He quoted

Michon, Gergoire and Lafont (1935) as stating that symptoms had been reported as intensified during menstrual periods and pregnancy.

The incidence of fractures with vertebral hemangiomas is infrequent. Holta (1942) described such a case in the fourth cervical vertebra of a sixty-six-year-old man with compression myelomalacia. On the other hand, Griep (1942) described a case of fracture of the eighth and twelfth dorsal vertebra and the second lumbar vertebra which were otherwise healthy, while the third lumbar vertebra was undamaged even though it showed characteristics of hemangioma.

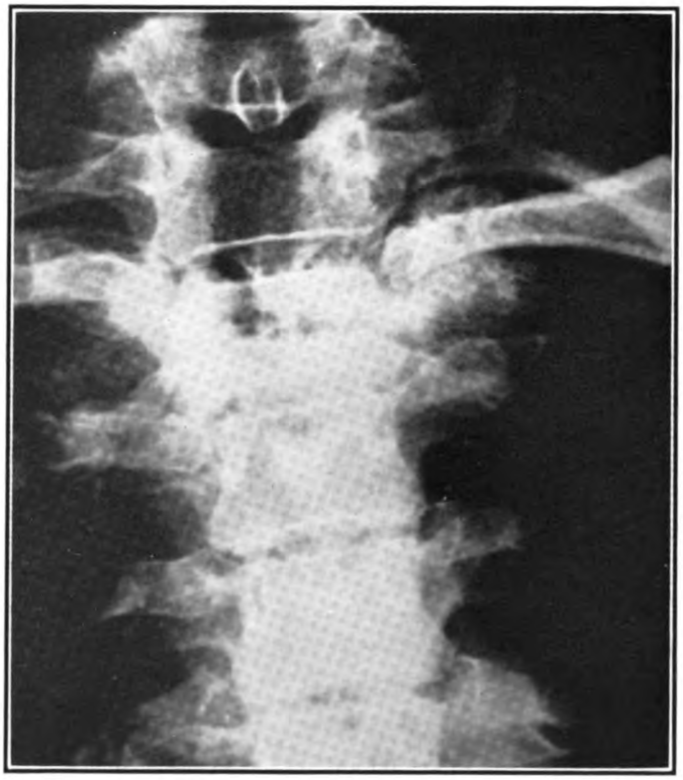

Fig. 237.—Hemangioma of second thoracic vertebra with left paravertebral extension of calcification within hemangiomatous tissue. Operative verification.

Asymptomatic vertebral hemangiomas which are the most frequent variety, require no treatment. However, in the event of pain without central nervous symptoms, *x*-ray therapy alone may prove efficient in relieving the patient's distress. When symptoms of central nervous system compression are only minimal, *x*-ray therapy may be cautiously attempted with the thought in mind that laminectomy may be required afterwards. However, in the event of rapidly progressive symptoms, decompressive laminectomy followed by *x*-ray therapy is the method of choice. There are numerous reports in the literature of the efficiency of *x*-ray therapy in relieving the distress incident to hemangiomas of the vertebra, including those of Bucy and Capp (1930), Ferber and Lampe (1942), Kaplan (1946), Foster and Heublein (1947), Schlezinger and Ungar (1939), Brøbeck (1950), Manning (1951) and Lindqvist (1951).

The plain film roentgen change seen most often with hemangiomas of the vertebra is a vertical, slightly wavy, parallel striated cancellous marking, most often seen in the vertebral body.

We have observed cases in which the vertical striations extended into the neural arches, and in 3 the changes were also present in the transverse and spinous processes. In 1 unusual case of a hemangioma of the first thoracic vertebra (Fig. 237) the patient was paraplegic. The vertical striations in this case were more or less obliterated by a peculiar overlying increased density. Lateral to the vertebral body was a smoothly lobulated tumor mass made up of fairly dense tissue. At first it was believed that we were dealing with an osteochondromatous sarcoma with spinal cord compression. Myelography in this patient disclosed a complete block, and a laminectomy for relief of the spinal cord compression was done. The tumor tissue removed was characteristic of vertebral hemangioma, and the patient did well following decompression and *x*-ray therapy.

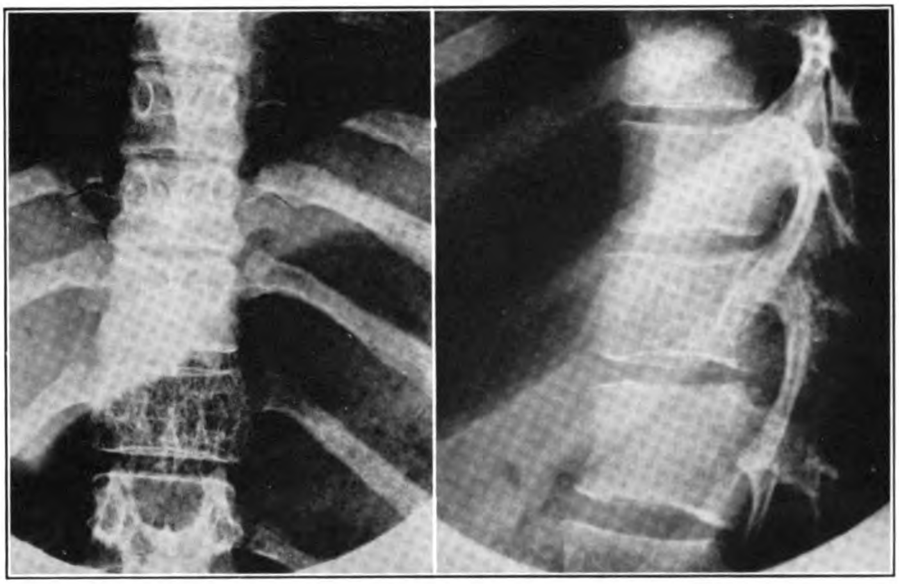

FIG. 238.—Hemangioma of the twelfth thoracic vertebra in a 23-year-old pregnant woman who became paraplegic at about the seventh month of pregnancy after having complained of back pain for about five months. Operative verification of spinal cord compression at laminectomy.

This case points out that hemangiomas may perforate through the cortex of the vertebral body peripherally and extend into the adjacent soft tissues. By far the more common method of progression of the tumor, however, is extension into the spinal canal.

Myelographic examination is indicated whenever there is a question as to whether or not the symptoms are secondary to compression myelopathy. In 3 cases observed by us a complete block was encountered, an observation reported before by several of the observers mentioned previously in this section. Lindqvist (1951) mentioned that in his case a relative block was noted in the anterior aspect of the spinal canal at the level of the ninth dorsal vertebra, but a contrast fluid was able to pass the obstacle in a thin band 3 mm. in breadth.

One other case observed by us appears to be worthy of record inasmuch as it appears to confirm the observation of Michon, Gregoire and Lafont (1935). This patient was a nineteen-year-old girl, who during the third trimester of pregnancy, suddenly began to complain of pain in the back followed by progressive weakness of the lower extremities which soon progressed to a paraplegia. In view of the urgency of the situation, laminectomy was done the day after demonstration of a hemangiomatous lesion involving the neural arches as well as the body of the twelfth dorsal verbetra (Fig. 234). This was followed by slow improvement, and one month later, when the obstetrician judged the time propitious, a cesarean section was done and a normal infant delivered. *X*-ray therapy was instituted immediately after delivery of the infant. Slow, progressive improvement followed, and the patient was symptom free six months later.

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MASSIVE OSTEOLYSIS

Decalcification of bone is a nonspecific phenomenon related to such diverse causes as infection, trauma, central nervous system disorders, metastatic tumors, metabolic conditions, the various reticuloses, as well as other processes. A remarkable, infrequent form of extensive osteoclasis is that associated with vascular changes of a benign nature with markedly prominent venous channels and without bone reaction. It has been attributed to a variant form of hemangioma in which the bone becomes weakened, soft and spongy, and filled with a benign proliferation of endothelial-lined channels which can be outlined radiologically after the injection of water soluble contrast material. The bone practically disappears progressively, giving rise to various appelations including phantom bone, vanishing bone, disappearing bone, cryptogenic osteolysis and essential osteolysis. The condition appears in patients from childhood to middle age, most often in children and young adults. There are no endocrine or chemical disturbances which might point to a causative mechanism. Complaints are referable to pathologic fractures with pain, swelling and deformities predominating. The lesion can originate in almost any bone except the skull and phalanges, mostly in the pelvis, sacrum and shoulder girdles. The osteolytic process sometimes ceases spontaneously, but reossification does not take place. The spine is rarely involved. In a few the disease is progressive and fatal in about two years.

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OSTEOID OSTEOMA

This lesion was designated by Jaffe (1935) as a benign osteoblastic tumor composed of osteoid tissue and atypical bone. As first reported, the tumor involved spongy bone. In 1940 Jaffe and Lichtenstein observed the same lesion in the cortex of the shafts of long bones accompanied by a considerable degree of periosteal reaction. Pathologically the lesion is characterized by the presence of a nidus, which on cut sections stands out from the surrounding bony tissue as a slightly bulging, rather rubbery, circumscribed red nodule surrounded by thickened and sclerotic bone. This nidus is the osteoid osteoma itself.

An osteoid osteoma within spongy bone merely provokes a thin rim of sclerotic reaction, quite different from the rather pronounced periosteal proliferation of bone observed with the same lesion developing in the cortex.

The lesion is one which affects relatively young people, from childhood to early adult life. It is accompanied by a considerable degree of pain which often is more severe at night. Of interest is the fact that the pain seems to respond quite readily to aspirin. Sharp tenderness over the involved area may be elicited on physical examination.

The usual sites of origin of these benign tumors are in the shafts of the long bones, particularly the femurs and tibias. Lesions have also been identified in other long bones and in the feet. Occasional instances of involvement of the humeri have been reported. Osteoid osteomas may appear in the vertebral column, and usually involve the neural arches or the articular facets. In a review of 62 cases the vertebræ were involved in

4 (Jaffe, 1945). In this paper Jaffe presented a roentgenogram showing an osteoid osteoma on the right side of the isthmus between the ascending and descending articular facets of the second lumbar vertebra. In a photomicrograph from another case of osteoid osteoma also in an articular facet of the second lumbar vertebra, Jaffe showed how the osteoid osteoma was sharply demarcated from the unaffected bone area.

Kleinberg (1944) reported a ten-year-old girl who had been treated for about a year for backache attributed to strain, scoliosis and osteochondritis. Lateral roentgenograms showed a slight kyphosis at the level of the first and second lumbar vertebra, with diminution in the size of the intervertebral disc. A diagnosis of possible Pott's disease was made. Pressure over this area produced pain. Later films showed an excavation and absorption of part of the right lamina of the second lumbar vertebra, which had been previously overlooked. At operation a red granular mass the size of a cherry was found between the laminæ of the first and second lumbar vertebræ on the right side, with erosion of part of the lamina of the second lumbar vertebra. Complete recovery followed excision of this lesion, which proved to be an osteoid osteoma.

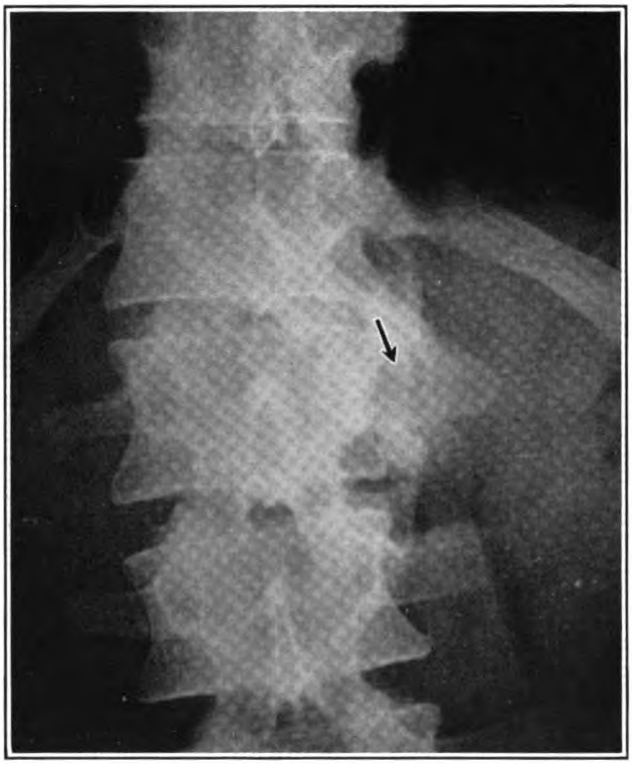

FIG. 239.—Osteoid osteoma of the left transverse process of the first lumbar vertebra. Considerable bony reaction is present. A small central densely calcified core is seen within a radiolucent focus (arrow).

Lewis (1944) included a case in which a nidus with surrounding sclerotic reactive bone tissue was encountered in a small articulation between the tenth and eleventh dorsal vertebra in a fifteen-year-old male. Sherman

(1947) mentioned that 128 reported cases of osteoid osteoma were available in the literature, 11 of which were in the vertebræ. In his series 7 additional cases were presented, making a total of 18 cases. An additional case of involvement of the right lamina of the third lumbar vertebra with clinical manifestations suggesting protrusion of an intervertebral disc was described by Jackson, Dockerty and Ghormley (1949). Two years later Dockerty, Ghormley and Jackson reported a clinical pathologic study of 20 cases in which the vertebræ were involved in 5. In 1 case the transverse process of a vertebra was involved destroying the entire bony structure with no surrounding sclerosis of bone. Of interest is the fact that these authors mentioned a case in which there were typical clinical and *x*-ray findings, and in which successive roentgenograms demonstrated eventual disappearance of the lesion without surgical intervention. Sabanas, Bickel and Moe (1956) also observed spontaneous disappearance in three cases observed from four to eight years.

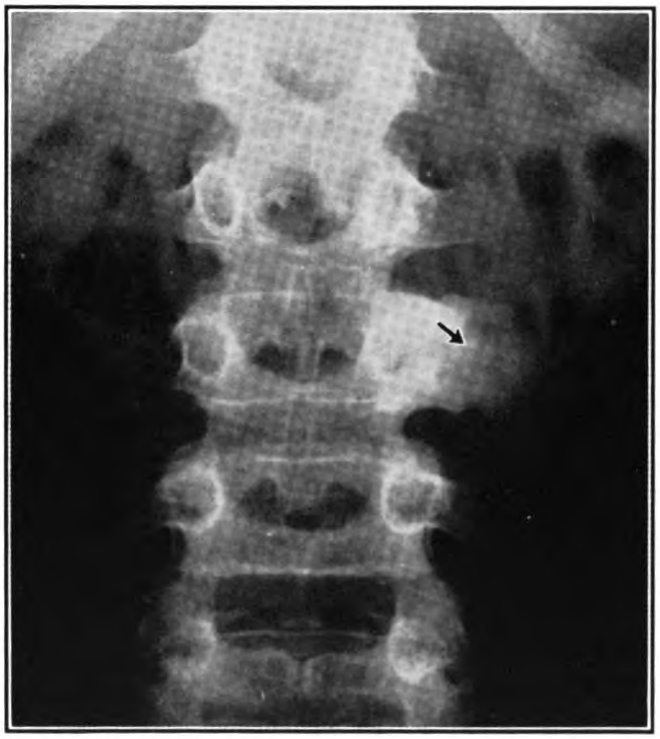

FIG. 240.—Osteoid osteoma of the left transverse process of the third lumbar vertebra. Note the heavy bony reaction, and the small calcified central core within a radiolucent circle (arrow).

It is apparent that the chief sites of vertebral involvement with osteoid osteoma are the neural arches, particularly the isthmus and the articular facets, and the transverse processes. Walker (1952) in a description of 3 cases of osteoid osteoma in children from nine to fourteen years of age, mentioned that the spinous process, pedicle and body were involved in 2 of these patients. One of these was of particular interest in that the spinous process of a fifth cervical vertebra had been involved with relatively little surrounding sclerosis of bone. The second case showed involvement of a pedicle and the body of the fifth cervical vertebra. This is the only case of involvement of a vertebral body which I encountered in the literature.

The radiologic diagnosis of osteoid osteoma insofar as the vertebræ are concerned is dependent upon the identification of the nidus. As a rule, the degree of sclerotic reaction around the nidus is slight, although in 1 case of an osteoid osteoma in the pedicle of the second lumbar vertebra seen by us a considerable degree of bony reaction could be identified radiologically as well as at subsequent operation. As a rule, the nidus is circular or oval in configuration and rarely exceeds 1 cc. in diameter.

An interesting roentgenogram is shown by Lichtenstein (1952) in which an osteoid osteoma is seen in the inferior facet of the fifth lumbar vertebra, the shadow being identified by a circular radiolucent area within which an island of bone of relatively normal density was present. A sciatic syndrome, which had been present clinically for over two years, was completely relieved after removal of the osteoid osteoma.

The outstanding change produced by osteoid osteoma is a radiolucent focus of osteoid which becomes opaque when calcification or ossification appears within it. The lesion then appears as a central circular area of increased density surrounded by a radiolucent halo. This, in turn, often is enveloped in the sclerosing reactive bone which varies considerably in extent, particularly in the vertebræ. The differential diagnosis between osteoid osteoma and sclerosing osteomyelitis is facilitated by the recognition of the nidus and halo, but it is quite possible for a small central rarefied focus to be present in an inflammatory lesion. In such cases biopsy is required. Removal of the lesion usually results in cessation of symptoms. However, in the spine the problem is somewhat less difficult because the sclerosing osteitis is not as frequent. Body section radiography is helpful in the investigation of suspected osteoid osteomas of the neural arches and its processes.

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BENIGN OSTEOBLASTOMA

This benign bone tumor resembles osteoid osteoma histologically in that it contains heavily vascularized osteoid and bone forming tissue. It occurs most often in patients from 10 to 25 years of age, but has been observed in younger and older individuals. It differs from osteoid osteoma in that pain is less intense. The tumor is considerably larger and grows rapidly. It has a predilection for the neural arches and pedicles of the vertebræ, and may cause compression on the spinal canal and intervertebral foramina. It also is seen in tubular bones, including those of the hands and feet, where they often are eccentric in position. Occasionally benign osteoblastomas occur in the ribs and skull.

The radiologic picture is not necessarily characteristic. The nidus seen with osteoid osteoma is absent. In the main, the lesion is lytic, produces expansion of the involved bone, thinning and eroding the cortex but remaining well circumscribed in a calcific shell. In some a sclerosing, stippled deposition of bone is present, which may help differentiate the lesion from other bone new growths.

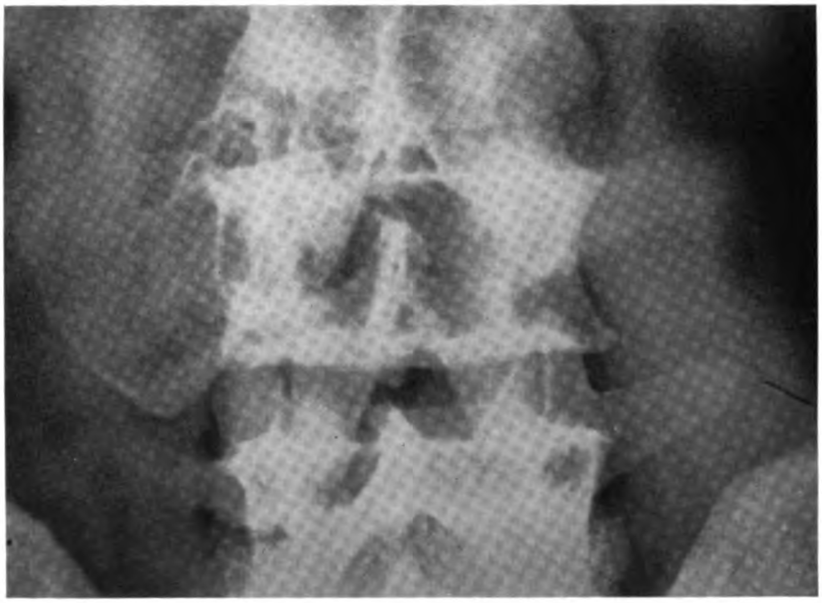

FIG. 241.—Benign osteoblastoma involving the right pedicle and transverse process of the fourth lumbar vertebra, eroding the right lateral border of the body.

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NASOPHARYNGEAL NEOPLASMS

It is generally agreed that most nasopharyngeal tumors exclusive of those arising as primary or metastatic growth in the subjacent bone originate in the soft tissues of the nasopharynx. Among these tumors one may encounter lympho-epitheliomas, transitional and squamous cell carcinomas, lymphoblastomas and sarcomas. As a rule these tumors manifest themselves first by proliferation in the soft tissue of the nasopharynx and then by extension into the base of the skull, usually by means of the basilar foramina. Involvement of the cervical spine by such tumors is rare. Such a case was reported by Geist and Portmann (1952) who described destruction of the first and second vertebral bodies by a nasopharyngeal malignancy. The roentgenograms of this patient showed an area of decalcification involving the anterior aspect of these vertebral bodies. Occasionally a nasopharyngeal tumor may be identified on *x*-ray examination of the cervical spine because of soft tissue swelling in the upper prevertebral area, best seen on lateral projections. As a rule this occurs only when the tumor is larger and has burst through the nasopharyngeal confines.

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Chapter

6

Spinal Traumatic Changes

FRACTURES AND DISLOCATIONS OF THE CERVICAL SPINE

Industrial, automobile and sports accidents are the most frequent causes of injury to the cervical spine. The vertebral bodies, the intervertebral discs and the contents of the spinal canal are affected by the trauma depending on its direction, force, duration and the posture and muscular tone of the patient at the time of injury. Often the adjacent soft tissues, the muscular and ligamentous structures, are severely injured, but these are difficult to evaluate in the absence of skeletal changes. With displacement of various components of the vertebra and intervertebral disc, acute or chronic spinal cord or nerve root compression supervenes, followed by a clinical picture in accord with the compressive phenomena. These vary from immediate transection of the cord to the results of more or less gradual compression of the anterior aspect of the cord. With certain traumas the central area of the cord is injured, while with others the anterior aspect of the cord or the nerve roots are disturbed.

In a review of 59 cases of cervical spine trauma, Crooks and Birkett (1944) found six fractures of the sixth or seventh cervical spinous processes or both. One fracture of the transverse process of the seventh cervical vertebra was seen. Fracture or fracture dislocation of the atlanto-axial region occurred in three, and there were two with displacement of the atlas on the axis. Four fractures of the dens were seen, with forward displacement in three and backwards displacement in one. The second cervical vertebra was fractured or displaced on the third in six cases. There were seven compression fractures, and ten fracture dislocations and subluxations. In eight cases there were dislocations with locking of the articular facets, and in five of these complete paraplegia appeared immediately after the injury. In a review of 87 lesions in 77 patients Rogers (1957) found anterior dislocation and bursting fractures of the vertebral body in 29 and ten patients respectively. Anterior fracture dislocation was present in eight cases, fracture of the dens in nine and six had wedge-compression fractures.

Injuries of the cervical spine have been classified according to the forces involved, the resultant displacements, and compressive or disruptive changes. Anterior dislocation, with and without fracture, occurs when there is a shearing force from behind which produces anterior displacement of the affected vertebræ. The posterior segments of the displaced vertebræ are tipped anteriorly and cephalad by the altered inclination of the articular facets. Dislocation is complete when the inferior articular process of the displaced vertebra becomes anterior to the superior articular process of the vertebra. If the displacement is such that the inferior articular process of the involved vertebra remains posterior, but is displaced anteriorly and upwards, partial dislocation is present. Fractures occur when longitudinal compression is added to this type of force. Compression of the veretbral body or disruption of the pedicles or laminæ add considerably to the possibility of concomitant cord injuries.

Posterior dislocation of the vertebra results when the shearing force is applied anteriorly, as in a fall on the face. The anterior spinal ligaments are are torn and the intervertebral disc is disrupted. The ligamenta flava and interspinal ligaments partake in the injury, and the spinal cord and nerve roots may be injured severely. With this type of injury the elements involved may reassume a normal relationship, leaving a lacerated spinal cord.

The most frequent cause of injury of the first and second cervical vertebræ is a blow on the vertex of the head or a fall from a height striking head first. The fracture results from pressure on the vertex and counterpressure from the spine squeezing the lateral masses of the atlas between the occipital condyles and the axis. The posterior and anterior arches are the weakest points of the axis, and fractures through these result in lateral displacements of the lateral masses. Fractures also occur through the grooves for the vertebral arteries. Unilateral rotary dislocations follow forced lateral flexion and rotation away from the dislocated fragment. The weakest point of the dens is its base. A force exerted from behind will result in a forward dislocation, while if the force is applied to the forehead with the head in hyperextension a posterior dislocation follows. The forward dislocation is more frequent, but paralysis is observed more often with backward displacement.

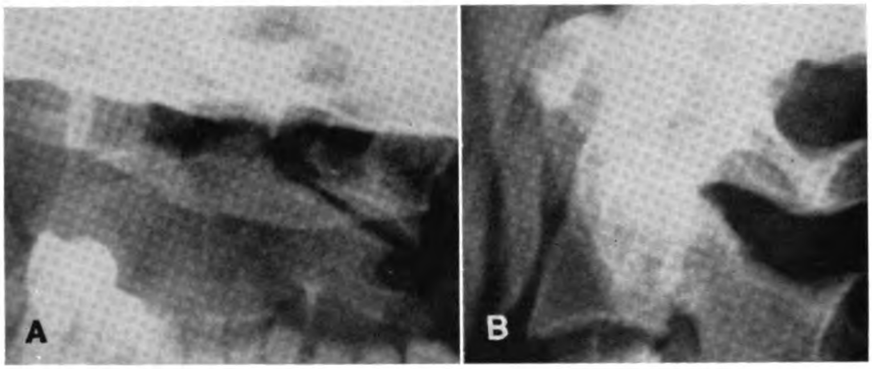

FIG. 242.—*A*, Anteroposterior view of fracture dislocation of first and second cervical vertebræ. The dens is fractured through its base, and there is displacement to the left of the lateral masses of the first cervical vertebra. *B*, Same case, lateral view. The space between the dens and the anterior arch of the first cervical vertebra has been increased.

Careful neurologic examination should precede radiographic examination of the cervical spine to avoid endangering the patient. It is advisable to review the preliminary films of the cervical spine before proceding with more complete examinations. Our first films are made in the anteroposterior, lateral and open-mouth projections. If indicated, flexion and extension lateral studies, stereo oblique views and body sections examinations are made.

Other mechanisms for fracture of the atlas include extreme extension, causing a fracture at the weakest point of the posterior arch close to the lateral masses on one or both sides. Pressure of the odontoid against the anterior arch may result in a fracture of this area. There may be dislocation of the lateral masses towards the head in bilateral fractures of the posterior arch because of upward pulling of the rectus capitis posterior minor muscle. Plaut (1930) included a table reporting 93 fractures of the atlas as gathered from the literature and mentioned that an overwhelming majority recovered to full occupational activity.

Fractures and fracture dislocations of the second cervical vertebra occur quite frequently as a result of trauma incident to accidents such as diving injuries which produce direct trauma on the occiput of the skull associated with a torsional force as the body swings over to the side when the head strikes the bottom. In these cases there may well be a lateral displacement of the atlas on the axis together with a fracture through the base of the odontoid process. An unusual case of this variety was reported by Davies (1945) in a thirty-eight-year-old man who had fractured his first and second cervical vertebræ after diving into a swimming pool twenty-three years before admission to the hospital. An ununited fracture resulted at the base of the odontoid, with a false articulation. Movement occurred at this false joint which subjected the patient to attacks of unconsciousness, which the author attributed to compression of the vertebral arteries, whose course he postulated was tortuous and subject to compression following a blow or twist of the neck.

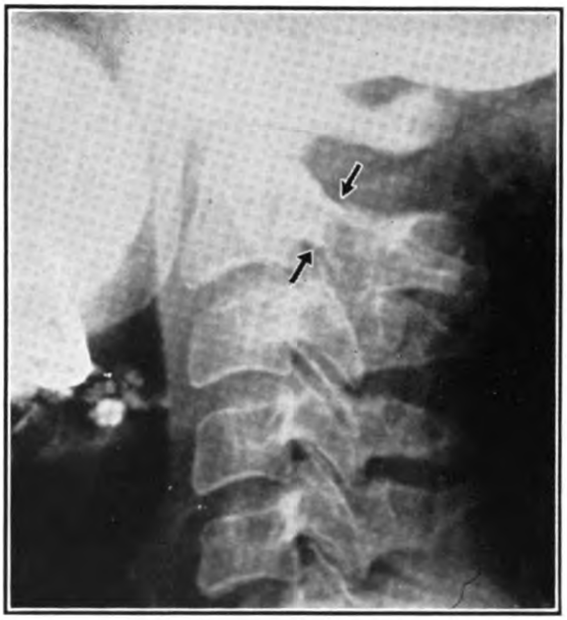

FIG. 243.—Fracture through the base of the spinous process of the second cervical vertebra (arrows).

In a case seen by me (Fig. 242) the patient was struck with the butt of a pistol during a holdup, and he fell heavily to the floor twisting his head. There was a fracture of the base of the odontoid process with a lateral shift of the dens as well as of the lateral masses of the first cervical vertebra towards the left side. In the lateral projection the space between the anterior arch of the atlas and the odontoid was increased to 1 cm. This patient had complained of pain on motion of the neck, but the condition remained undiagnosed for about two weeks before he came for medical attention. There were no central nervous system symptoms and he responded well to conservative treatment. Another patient with a similar type of injury had a fracture through the base of the spinous process of the second cervical vertebra (Fig. 243).

Malformations of the occipitocervical junction, when present, influence the type of fracture. With occipitalization of the atlas or basilar invagination the dens is high in the spinal canal in relation to the malformed foramen magnum. Therefore the dens is more susceptible to injury as being so situated as to be more likely to injure the cord and the medulla oblangata.

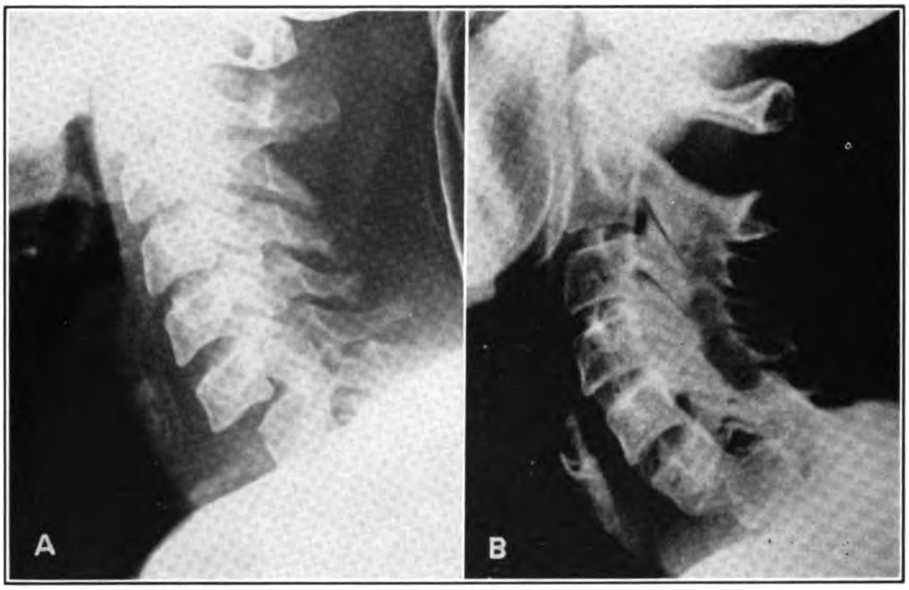

FIG. 244.—*A*, Hyperflexion fracture dislocation of the sixth on the seventh cervical vertebra due to disruption of the neural arch. *B*, Old hyperflexion fracture of lower cervical spine due to automobile accident seventeen years before. Fusion has developed between the sixth and seventh cervical vertebræ. The patient became asymptomatic after prolonged treatment.

In young people the dens may separate at its basilar epiphysis. One should not interpret a normal persistent epiphyseal line as a fracture.

Neurological sequelæ following fracture of the dens occur immediately or later. Hemorrhage into the spinal canal or outside the canal with disruption of adjacent nerves, contusions of the greater occipital, glossopharyngeal, palatal or chorda tympani nerves may occur. Immediate paralysis below the lesion, hypalgesia and hypesthesia together with preserved position, touch, motion and vibratory sense after injury may indicate the necessity for surgical relief. Myelography is contraindicated under these circumstances. Schneider regards these symptoms as the direct effect of trauma on the cord rather than vascular impairment due to injury to the anterior spinal artery.

Delayed consequences of high cervical trauma include failure of healing of the fracture, with persistent abnormal mobility, excessive callus formation at the fracture site or necrosis of the dens. Compressive changes of the anterior aspect of the spinal cord are important, and simulate various degenerative diseases with progressive weakness, spasticity, hyperreflexia and absence of sensory disturbances. Pain and numbness of the lower extremities can be produced by lower cervical spinal injuries, with no symptoms directly referable to that area, and in the absence of lumbar discal changes. The situation becomes confused when minor or no changes are found on lumbar myelography, and the cervical canal should always be studied carefully in such cases.

Injuries to the lower cervical spine frequently involve the bodies of the fourth, fifth, sixth and seventh cervical vertebræ. Many of these follow a so-called "whiplash" injury which causes hyperflexion followed by a spontaneous extensor recoil (Davis, 1945). Head-on automobile collisions, or sudden stops, are common causes. This may result in a partial compression of the anterior aspects of the lower cervical vertebral bodies. Injuries to the transverse and spinous processes, the laminæ and pedicles may also occur as a result of these injuries, together with subluxations of the articular facets. The clinical picture varies greatly, depending on the location and extent of the injury. These may be very difficult to identify, and one encounters a considerable disparity between so-called negative roentgenograms of the neck and a patient who is obviously and painfully injured. In these cases the usual lordotic curve of the cervical spine is straightened, an observation which should induce further detailed examinations, particularly stereoscopic oblique studies. A so-called "sprain" may well be a subluxation of an articular facet almost indistinguishable even on the most complete roentgenographic examination. In such cases stereoscopic lateral studies in flexion and extension, as well as oblique stereoroentgenograms are essential. Slight changes in the alignment of the articular facets, increased spacing between the spinous processes and minor alterations in the configurations of the apophyseal joints point towards minor dislocations which may be asymptomatic or productive of considerable discomfort. These are difficult to evaluate and often give rise to much discussion, particularly in medicolegal situations.

In injuries of the cervical spine in which the intervertebral foramina are affected, one may encounter a syndrome simulating that of a ruptured cervical intervertebral disc. Two such cases were reported by Kristoff and Dratz (1948) who observed that fracture of the margins of an intervertebral foramen resulted in compression of the corresponding nerve root, with pain identical to that seen with herniated discs. The pain may be radicular and aggravated by motion of the head. They believed that myelography may be unnecessary if the plain films are carefully made, and that myelograms may or may not show a filling defect with such injuries.

The dramatic effect of hyperflexion injury upon the intervertebral discs of the lower cervical spine has been the subject of several reports. Brooke (1944) reported a case of quadriplegia with complete transverse cervical myelitis produced by traumatic herniation of an ossified nucleus pulposus. At necropsy fifteen days after admission, a defective and frayed posterior longitudinal ligament was found, with posterior protrusion of a calcified nucleus pulposus between the fourth and fifth cervical vertebræ. The cord at this level was softened. In this patient, who was a fifty-six-year-old male, the plain films revealed narrow interspaces between the fourth and fifth and fifth and sixth cervical vertebræ, but the vertebræ were normally aligned. A soft defect was palpated in the spinal cord opposite the interspace between the fourth and fifth cervical vertebræ, and directly anterior to this was a hard, irregular calcified intervertebral disc which measured 1 × 1.5 × 0.5 cm. On slight flexion of the head this bulged about 1 cm. into the spinal canal. It appeared obvious that the herniated disc was responsible for the area of cord softening. The disc itself was readily lifted from its bed, and could be completely removed after cutting a few fibers of the posterior longitudinal ligament.

Cramer and McGowan (1944) pointed out that recoil injuries with extreme retroflexion may cause similar damage to the spinal cord by posterior displacement of the nucleus pulposus in the lower cervical spine. They described a case in which the cervical cord of a patient who died after a diving injury was almost completely severed anteriorly, without dislocation of the spinal column. Necropsy revealed that the intervertebral disc had protruded directly opposite the lesion in the spinal cord, and they postulated that violent protrusion of the nucleus pulposus due to compression of the intervertebral disc was the mechanism for the spinal cord injury in their patient. These authors did not believe that the explanation could be an extreme vertebral dislocation with immediate spontaneous reduction. This possibility was excluded because the construction of the cervical spine is such that dislocation occurring under great stress must be associated with tearing of the ligaments, fractures of the bones, or both conditions.

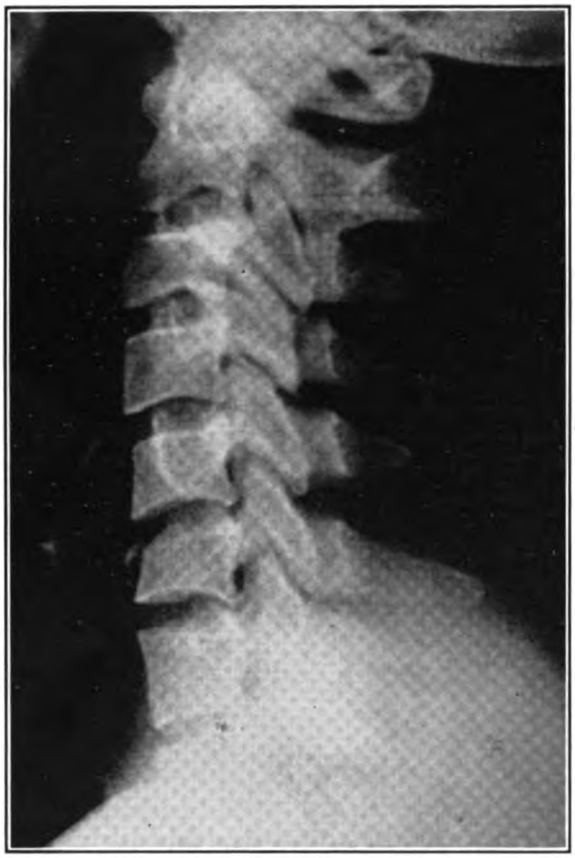

FIG. 245.—Chip "tear-drop" fracture of anteropsuperior aspect of the body of the sixth cervical vertebra, with loss of cervical lordotic curve due to intense muscle spasm. This was caused by a diving accident.

Another case of paraplegia following a hyperextension cervical spine injury was reported by Taylor and Blackwood (1948) who indicated that myelography may be required to demonstrate prolapse of the disc, particularly if the plain film roentgenograms of the cervical spine were normal. In their opinion, spinal canal block may develop later by reason of edema of the cord due to contusion, because of protrusion of a nucleus puplosus, or dislocation with immediate spontaneous reduction.

A most unusual case of fatal nucleus pulposus embolism involving a large segment of the spinal cord and medulla was reported by Naiman, Donohue and Prichard (1961) in a 15-year-old boy who died shortly after a minor injury with progressive quadriplegia, respiratory paralysis and sensory loss.

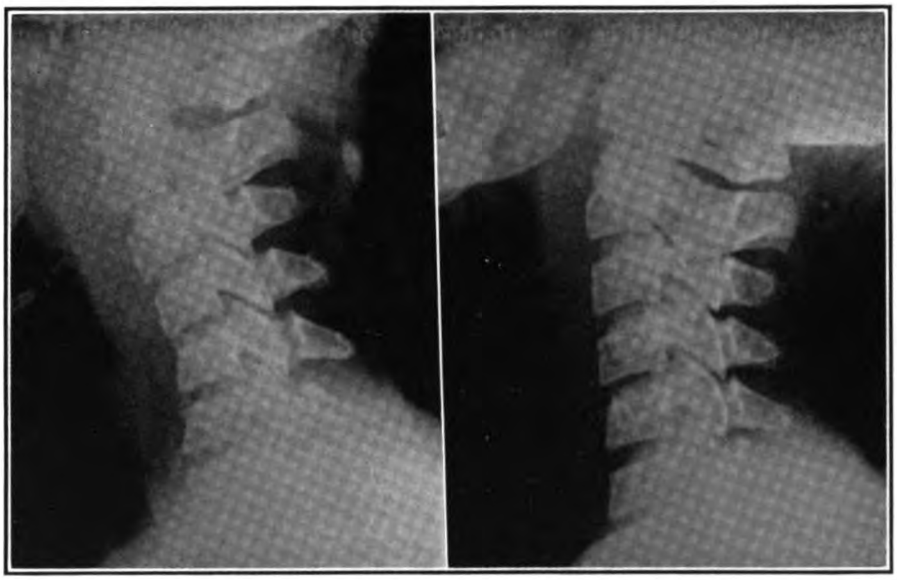

FIG. 246.—Reversal and straightening of the normal lordotic curve following hyperflexion injury. Films made in attempted flexion and extension. No bony injury could be identified on the oblique or frontal views as well as on the lateral films shown here.

The role of acute herniation of the nucleus pulposus of the lower cervical spine in the production of cord injuries was described by Schneider (1951). He maintained that an immediate complete paralysis with hyperesthesia at the level of the lesion, and an associated sparing of touch and some vibration sense constituted a syndrome for which early operation was necessary. This author also stressed the role of the taut dentate ligaments which hold the cord relatively firmly in place, thereby enhancing the traumatic effect of protrusion of the nucleus pulposus. He believed that even though no anatomical extrusion of disc material was present, immediate decompression by laminectomy, incision of the dura and sectioning of the dentate ligaments bilaterally might relieve pressure. Myelographic examination of these cases may show spinal canal block, confirming the need for operation.

Delayed paraplegia following injuries to the vertebræ was mentioned by Rogers (1945) who reported 4 cases in which paraplegia developed from a few minutes to forty-eight hours after injury. All were able to walk during this interval. The effect of injuries to the cervical spines of elderly individuals with arthritis was discussed by Barnes (1948). In these patients there is a tendency towards kyphosis of the thoracic spine, usually compensated for by a cervical lordosis. This results in a relatively fixed position of the cervical spine, and any injury may cause a degree of hyperextension because of the inability of the patient to flex his neck. Under such circumstances strain develops on the anterior spinal ligament, which may rupture and be accompanied by a chip fracture of the vertebral body. The intervertebral disc may be torn, but there is little chance for a posterior displacement of the nucleus pulposus under such circumstances. Kaplan (1953) postulated that hyperextension injuries might cause thrombosis of the spinal arteries with consequent cord damage, and reported a verified case in a fifty-four-year-old male who had been overthrown by a wave while sea-bathing.

Hyperextension injuries may produce serious damage in newborn infants, especially during breech deliveries. Leventhal (1960) reported these at the cervicothoracic junction, with injury to the brachial plexus. The spine itself is not damaged, but injury to the head and first cervical vertebra may occur. The development of meningeal adhesions might cause block as in two cases reported by him.

In evaluating the effects of hyperextension injuries of the cervical spine, the role of the ligamenta flava and the presence of spondylotic ridges should be considered. During the moment of extreme hyperextension the combination of wrinkling of the ligamenta flava anteriorly and the increased dorsad bulging of spondylotic ridges combine to cause marked compression of the cord. This is enhanced when the diameter of the spinal canal is reduced either as a developmental change consequent to increased thickness of the neural arches or because of associated hypertrophic spondylosis.

Some hyperextension injuries damage the central area of the cord, but more often the anterior aspect is affected. The role of the dentate ligaments in preventing dorsal protrusion of the cord against the neural arches enhances the probability for injury to its anterior aspect. It is noteworthy that considerable time may elapse before these injuries manifest themselves clinically, and their association with previous trauma sometimes is overlooked. Schneider and Knighton pointed out that the syndrome of chronic injury to the central zone is characterized by weakness of the fingers, hands and arms to a greater extent than seen in the lower extremities. The sensory pattern is erratic, and the entire picture easily confused with syringomyelia. The syndrome of acute injury to the central zone also includes the effects of damage in the vicinity of the central canal, with involvement of the nerve supply to the neck, shoulders and upper extremities and lesser damage to the nerves of the trunk and lower extremities. To the degree in which permanent damage exists, the chronic picture develops later. Considerable recovery may occur if the trauma results in a reversible form of injury.

Injuries to the spinous processes of the lower cervical spine constitute an interesting group of fractures which have been referred to as "clay-shoveler's" fractures. Such fractures may occur in one or several vertebræ, from the sixth cervical to the third thoracic segments. The fracture line usually appears at the narrowest part of the spinous process about 2 cm. from its end. According to Hall (1940) this type of injury is attributed to muscular pull caused by a sudden jerk while using a shovel. Annan (1945) felt that undernutrition and fatigue were contributing factors. The fracture is best seen on lateral roentgenograms. It may be identified in the anteroposterior view by a downward displacement of a tip of the spinous process as seen on an overexposed anteroposterior view (Zanca and Lodmell, 1951). With some hyperflexion injuries fractures of the spinous processes of the lower cervical and uppermost thoracic vertebræ may occur, resembling clay-shoveller's fracture. These tend not to heal, and may be followed by calcifications in the adjacent soft tissues due to the laying down of calcium in an accompanying hematoma (Gershon-Cohen, Budin and Glauser, 1954).

Vertical fractures of the cervical vertebral bodies were reported by Richman and Friedman (1954). All 17 cases in their series were in paraplegic patients, and were associated with subluxations or other cervical spine fractures. Laminagraphy was found of particular help in identifying these thin linear vertical fracture lines, which were observed more often in the lower cervical segments.

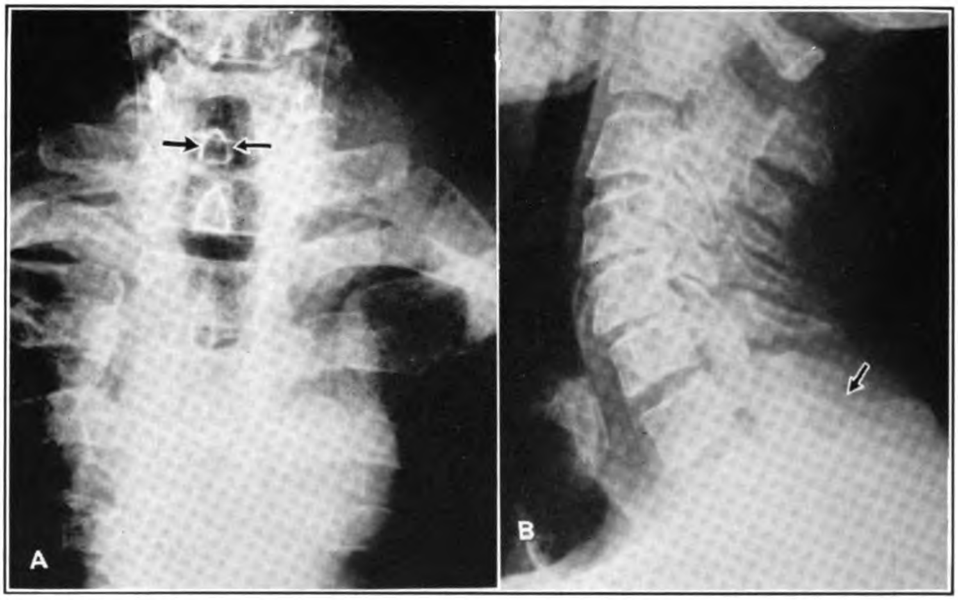

FIG. 247.—*A*, Fracture of spinous process of the seventh cervical vertebra. The anteroposterior view shows the tip displaced downwards (arrow). *B*, Same case, lateral view. The fracture is at the midportion of the spinous process, and the distal fragment is displaced downwards (arrows).

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Atlanto-Axial Dislocations

The normal atlanto-axial articulation consists of four joints, two pivotal and two arthrodial, or gliding joints. One pivot joint is between the posterior surface of the anterior arch of the atlas and the front of the odontoid process, and the second is between the anterior surface of the transverse ligament of the atlas and the posterior aspect of the odontoid process. The two gliding joints are those on either side of the articular processes between the atlas and the axis. On either side of the odontoid process is a bursa which separates it anteriorly from the transverse ligament of the atlas. Each of the four joints is lined with synovial membrane.

The structures connecting the atlas and axis consist of the articular capsules, which unite the margins of the lateral masses of the atlas with those of the posterior articular surface of the atlas. In front is the anterior atlanto-axial ligament, which extends from the inferior margin of the anterior arch of the atlas to the anterior aspect of the body of the axis. The posterior atlanto-axial ligament is attached from the lower border of the posterior arch of the atlas to the upper edge of the laminæ of the axis. The transverse ligament of the axis extends across the ring, keeping the odontoid process in contact with the anterior arch. Arising from either side of the upper portion of the odontoid process are two strong rounded ligaments called the alar ligaments, which insert into the occipital condyles on their medial aspects. These limit the motion of the head, and are referred to as check ligaments. Between the two alar ligaments lies the apical odontoid ligament, which extends from the tip of the odontoid process of the anterior margin of the foramen magnum, and unites with the deep portion of the anterior atlanto-occipital membrane and the superior crus of the transverse ligament of the atlas. This is regarded as a rudimentary intervertebral fibrocartilage, and traces of notochord may persist within it.

Radiologic investigations of the atlanto-occipital articulations should include films in the lateral and the anteroposterior positions, projecting the atlas and axis through the open mouth. In addition submento-vertical or vertical-submental views aid in visualizing the atlanto-occipital articulations. The latter views are sometimes difficult to obtain, particularly in patients with symptoms of atlanto-axial dislocation. An important aid to the radiologic examination of these joints is body section radiography, which brings out the articular surfaces between the occipital condyles and the first and second cervical vertebræ to a degree which cannot be obtained by routine methods. This examination is particularly important in those individuals who are unable to move their necks or open their mouths widely.

One must also keep in mind the wide range of variations in the normal configurations of the atlanto-occipital joints. Paul and Moir (1949) found considerable asymmetry in the lateral aspects of the articulations between the first and second cervical vertebræ. In a group of 5 individuals, 9 had some degree of lateral asymmetry. A distinct lateral motion of the atlas on the axis could be demonstrated on lateral flexion of the head, or by forceful pressure by the examiner against muscular resistance on the part of the patient. Such lateral mobility of the atlas in relationship to the axis had been noted before by Dankmeijer and Rothmeier (1943), who warned that it should not be mistaken for subluxation.

The configuration of the atlanto-axial joint in the lateral position was studied by Coutts (1934), who regarded the width of the joint space as an important index to its condition. The normal width of this joint is between 1 to 2 mm., and the normal upper range of the inclination of the atlas against the axis is 10 degrees. This angle is determined by a line joining the inferior borders of the anterior and posterior atlantic arches, and a second line joining the inferior border of the articular processes of the axis with the inferior border of its spinous process. Jackson, (1950) in a study of the distance between the anterior aspect of the odontoid and the posterior articular surface of the atlas, measured the range of distance between these two surfaces in flexion and extension in 50 normal individuals. He found that in children this space varied from 2 to 3 mm. and never exceeded 4.5 mm. In flexion or extension in adults the space never exceeded 2.5 mm., the measurement being greatest in flexion.

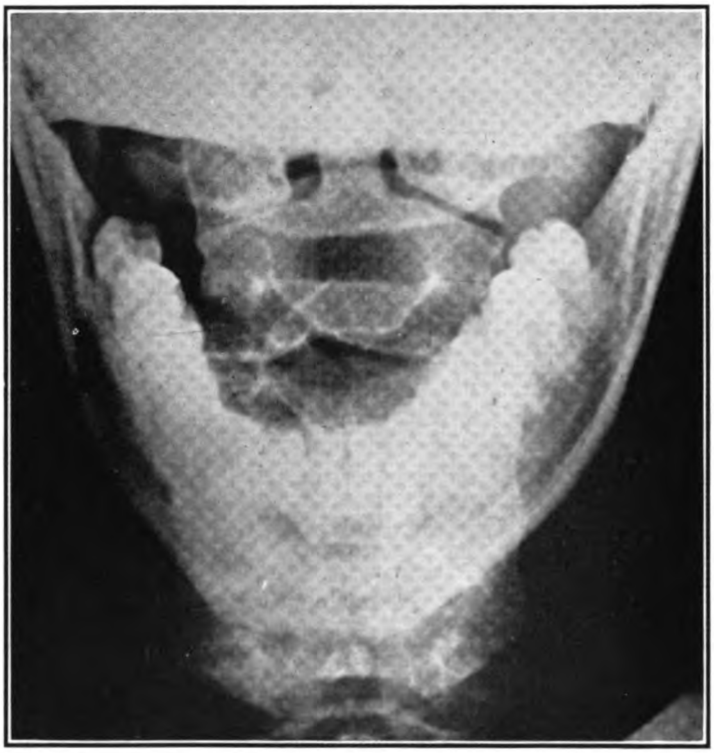

FIG. 248.—Atlanto-axial dislocation due to recent upper respiratory infection. The articulation between the right lateral mass of the first cervical vertebra and the underlying second cervical articular surface is markedly narrowed, while the left articulation is not visibly altered. The dens is closer to the left side.

The position in normal individuals of the tip of the spinous process of the first cervical vertebra and in patients suffering from spontaneous subluxations of the atlas was described by Hadley (1944). He noted that normally the tip of the first cervical spinous process was in a direct line with the corresponding structure of the second cervical vertebra. With spontaneous subluxation the tip of the first cervical spinous process moved forward. At the same time, the jaw is displaced forward from the spine, and the space between the odontoid and the arch of the atlas is increased. The arch of the atlas is moved anterior in relationship to the arches below.

Dislocations of the atlanto-axial joints are partial or complete. They occur either unilaterally, with anterior or posterior displacement of one of the lateral masses of the first on the second cervical vertebræ, or bilaterally with anterior displacement of the atlas on the axis. Rotational displacements also take place, with one lateral mass displaced anteriorly and the other posteriorly. There may be partial or complete loss of contact of the inferior articular surfaces of one or both lateral masses of the axis with the corresponding superior articular surfaces of the atlas. In addition, there may be right or left lateral dislocation of the atlas on the axis. In this circumstance a fracture of the odontoid process usually accompanies the lesion, and a history of trauma is generally obtained.

The etiologic factors concerned with dislocations of the atlanto-occipital articulation include trauma, congenital defects such as absence of the odontoid process or incomplete fusion of the odontoid process with the body of the second cervical vertebra (Clark, 1941; Weiler, 1942; Nievergelt, 1948), inflammatory diseases such as acute tonsillitis, cervical adenitis or the various exanthematous diseases (Sullivan, 1949). Cases have also been reported following mastoid infections (Frank, 1936), rheumatic fever (Evans, 1941), and after tonsillectomy (Swanberg, 1919). Tuberculosis, syphilis, or more often metastases, of the cervical spine, by destroying structures comprising the atlanto-occipital articulations, may be followed by dislocations following an unguarded sudden movement of the head or other trauma (Willard and Nicholson, 1941). Kyphoscoliosis in children might be a causative factor (Hess, Abelson and Bronstein, 1942).

Excluding those cases of obviously traumatic origin, those due to tuberculosis, syphilis, or other bone-destroying lesions such as osseous metastases, or to congenital anomalies of the atlas and axis, the etiology of spontaneous atlanto-axial subluxation has been ascribed by Wittek (1908) to an effusion into the joint capsule, with overdistention of the ligaments. Greig (1931) stated that with cervical infections a diffuse tissue hyperemia results, which produces decalcification of the adjacent bone and loosening of the intervertebral ligaments, particularly those between the atlas and axis. This may permit the skull and first cervical vertebra to dislocate anteriorly, or there may be a backward slipping of one articular facet of the atlas on the axis. Watson-Jones (1932) also ascribed the dislocation to hyperemia throughout the cervical tissues causing decalcification of the upper cervical vertebræ. This hyperemia is not accompanied by destruction of bone, and may be very difficult to identify roentgenologically. An inflammatory process anywhere in the upper cervical region may cause such decalcification, but inasmuch as the condition occurs mainly in children in whom the bones are imperfectly developed and not well calcified, roentgenographic identification of the demineralization may be lacking.

Frank (1936) compared dislocation of the atlas to subluxation of the hips, which sometimes complicates various exanthematous diseases, typhoid and mastoiditis. A case of an associated spontaneous dislocation of the hip with a spontaneous dislocation of the atlas was described by Steele (1937), who mentioned that distention of the joint and relaxation of the ligament was probably responsible for this change. Stein, Bloch and Kenin (1953) in a report of a case of a non-traumatic subluxation of the atlanto-axial articulations suggested that effusion into the atlanto-axial joint as a result of hypersensitivity might be a causative factor, comparing the change to that described by Edwards (1952) in transient synovitis of the hip joint in children.

Most cases of atlanto-axial dislocations occur in children. The disease usually appears about two weeks after the onset of an inflammatory lesion involving the nose and throat. The patient holds his head in a torticollis position, with the chin frequently pressed closely against the neck and pointed to the side opposite the dislocation if a unilateral dislocation exists. The patient may find it difficult to open his mouth and complains of pain in the occipital and mastoid areas. Some degree of flexion may be obtained, but extension of the head usually produces considerable pain. Rotation of the head from side to side is impaired, and the chin usually cannot be rotated beyond the midline. Considerable spasm of the posterior spinal muscles of the neck is present.

Central nervous system symptoms referable to atlanto-occipital dislocations are quite infrequent in cases of the spontaneous variety. Such a case, however, was reported by Wilson, Michele and Jacobson (1940) in an elderly patient who had pneumonia followed by atlanto-occipital dislocation resulting in quadriplegia. This patient recovered following treatment by head traction. Another case with nontraumatic atlanto-occipital dislocation accompanied by quadriplegia was reported by Titrud, McKinlay, Camp and Hewitt (1949) in a forty-year-old woman who recovered after

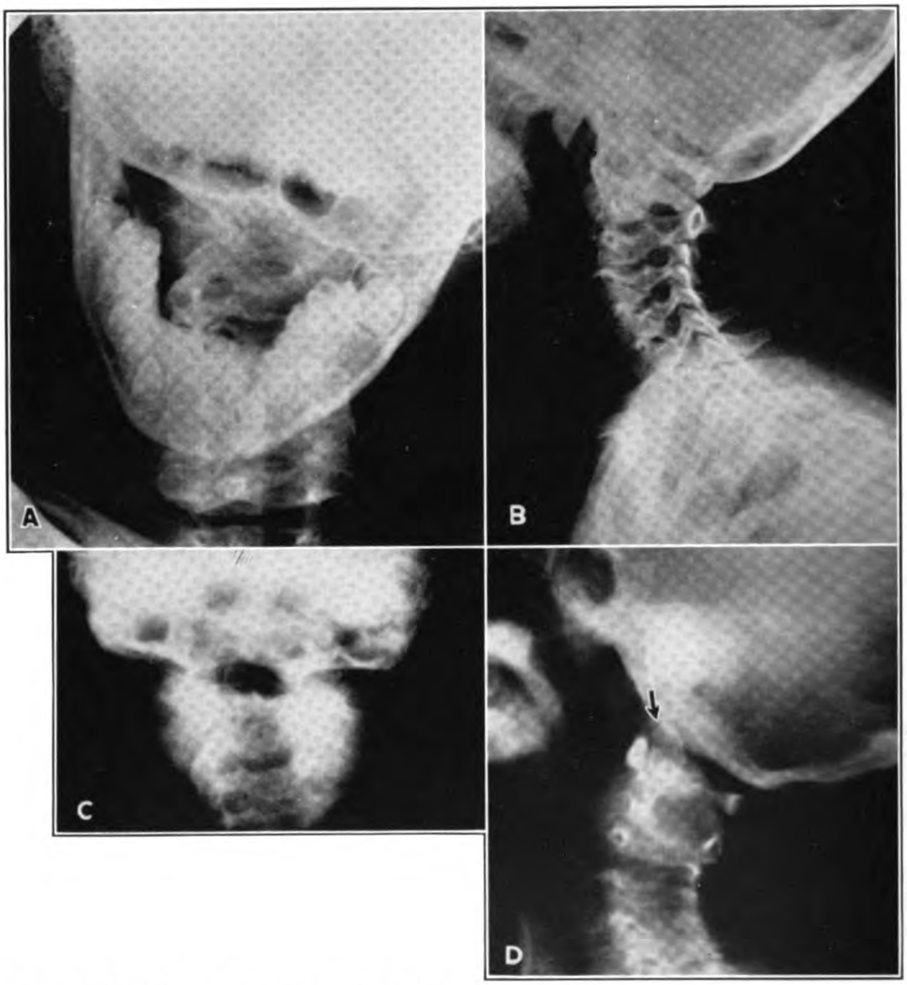

FIG. 249.—*A*, Atlanto-axial dislocation incident to a mild attack of measles. The head and jaw are tilted to the left. Note the tilting of the dens to the right. The left cervical mass of the first cervical vertebra is rotated out of view. *B*, Same case, lateral view. Because of the tilt of the head true laterals could not be obtained. *C*, Same case, anteroposterior body section roentgenogram. Note the tilt of the dens and the absence of the right lateral mass of the first cervical vertebra. *D*, Same case, lateral body section roentgenogram. The tip of the dens is well above the anterior arch of the first cervical vertebra (arrow).

surgical decompression followed by spinal fusion. Alexander, Masland and Harris (1953) reported an infant with anterior dislocation of the first cervical vertebra simulating cerebral birth injury. Myelography in this case revealed obstruction at the level of the first cervical interspace.

A review of our own cases of atlanto-occipital dislocation indicates that our findings have been much the same as reported by other observers. In 5 of our cases, however, we noted no gross forward displacement of the body of the atlas anteriorly, while the odontoid process was tilted dorsally. The views that were of greatest advantage were those made through the open mouth, and laminagraphic examination of these cases proved of considerable assistance by the elimination of the overlying shadows of the base of the skull and of the jaws. In such cases the asymmetry of the lateral masses of the first cervical vertebræ became obvious and the tilting of the odontoid process could be easily identified. It was noted that the odontoid process tilted opposite to the dislocated lateral mass and that the space between the odontoid and the undisplaced lateral mass appeared to be

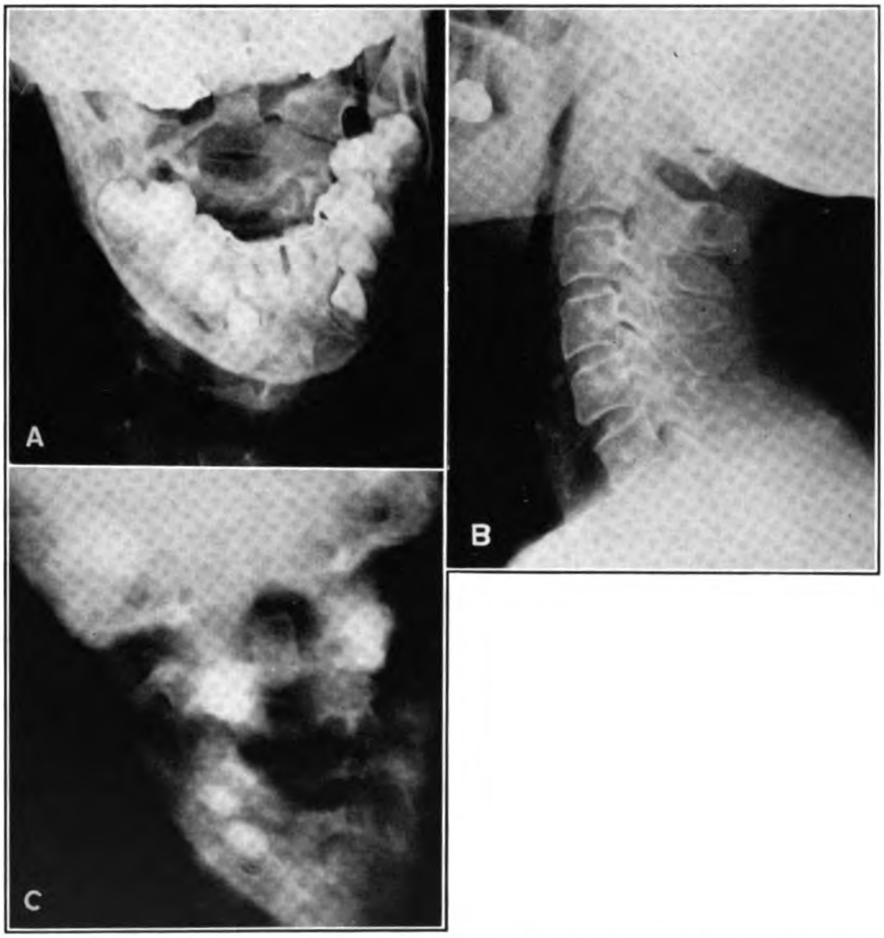

FIG. 250.—*A*, Atlanto-axial dislocation, with tilting of the jaw to the left and rotation of the right lateral mass of the first cervical vertebra out of view. *B*, Same case, lateral view. No displacement of the dens is seen. *C*, Same case. Anteroposterior body section roentgenogram.

narrowed. In 1 case (Fig. 250) the tip of the odontoid process could be beautifully demonstrated on a lateral roentgenogram and was displaced upwards so that it projected 0.5 cm. above the superior aspect of the anterior arch of the atlas.

It was also of interest that the space between the anterior aspect of the odontoid and the arch of the first cervical vertebra was within normal limits in 5 of the 8 cases observed.

Atlanto-axial dislocations were observed in 5 cases of metastases to the upper cervical vertebræ from carcinoma of the breast and 2 from bronchogenic carcinomas. In each a forward displacement of the head was observed, with loss of rotary motion and extension. Roentgenologic investigations disclosed destruction of the atlas or the axis or both, with disruption of the joints at the atlanto-occipital and atlanto-axial junctions. Forward displacement of the first or second cervical vertebral bodies was present in all 7 cases.

Another cause, mentioned in Chapter 4, of atlanto-axial dislocation is ankylosing spondylitis or rheumatoid arthritis. Colsen (1949) in reporting 2 such cases, mentioned that an injury may precipitate the dislocation. The occurrence of spontaneous atlanto-axial dislocation in ankylosing spondylitis and rheumatoid arthritis was magnificently reviewed by Sharp and Purser (1961). They noted that in patients with rheumatoid arthritis the gap between the dens and the anterior arch of the atlas might be considered as separated if it measured three millimeters, even though a measurement of up to four millimeters has been found in normal individuals. They reported a series of 22 patients with ankylosing spondylitis and 26 with rheumatoid arthritis with spontaneous atlanto-axial dislocation. In the spondylitic patients some clinical evidence of this displacement was found very frequently, but such evidence was absent in a fairly large number of patients with rheumatoid arthritis.

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FRACTURES OF THE THORACIC SPINE

Fractures of the thoracic vertebræ follow direct violence causing sudden severe hyperflexion, as may occur during an automobile accident or a fall. A similar injury may be incurred during athletic accidents, as in footfall or soccer. Infrequently, fractures of the upper dorsal spine occur if the patient is struck with a heavy object while the head is bent forward and the trunk is stooped anteriorly. Fractures of the dorsal spine usually are of the compression variety, with narrowing of the anterior aspect of the involved vertebral body. The most common site is the lower thoracic spine, but occasionally upper thoracic spine fractures are observed. Fractures in the middle thoracic spine are relatively uncommon. Fractures of the neural arches and of the spinous or transverse processes of the thoracic vertebræ likewise are infrequent. Those of the transverse processes of the seventh cervical or first thoracic vertebræ are very painful, and often are overlooked. Fractures of the thoracic vertebræ usually involve the superior aspects above the pedicles transversely. Compaction of cancellous bone results in some increase in the density of the injured vertebra. If the direction of the trauma includes a lateral and rotary force as well as forced flexion the resultant fracture produces lateral wedging in association with which injury to the ipsilateral pedicle and articular facet may occur. This is infrequent in the upper nine or ten thoracic vertebræ. The soft tissues also participate in the injury, with hemorrhage, laceration of various ligaments and resultant instability and dislocation of bony structures. The intervertebral disc may be torn. Hemorrhage into the spinal canal, displacement of bone or disc fragments, or edema consequent to trauma all can contribute to compressive spinal cord phenomena. Occasionally vertical fractures through the long axis of a vertebral body are seen, and less often the fracture involves only the lower aspect of a vertebra.

Simple minor fractures of a vertebral body usually are of little concern. When comminution of fracture fragments occurs, particularly with injuries of the neural arches and pedicles, or when dislocations are superimposed, serious difficulty may arise because of intrusion into the spinal canal. In such cases roentgenologic examinations must be detailed so that injuries to the smaller vertebral elements are visualized. Myelography may be required in addition.

Dislocations of the thoracic vertebræ are relatively uncommon and are usually seen with fractures. Stanger (1947) in a review of 43 patients, noted a relatively high incidence of paraplegia with such injuries to the thoraco-lumbar junction. His patients were coal miners who worked in a crouched position. In 30, the eleventh and twelfth dorsal and first lumbar vertebræ were involved. In our own experience there was but a single case with a forward displacement of the body of the tenth thoracic on the eleventh thoracic vertebra, together with fractures of the eleventh ribs and of the laminæ of the tenth dorsal vertebra. In addition to the forward displacement of the tenth on the eleventh dorsal vertebræ there was a right lateral displacement of approximately 0.5 cm. Interestingly enough, the intervertebral space was not altered, and there were no central nervous system complications. The trauma had been incurred during a head-on automobile collision. The patient had been seated in the front compartment, so that her chest was thrown violently forward while at the same time the lumbar area was forcibly displaced backwards, giving rise to a shearing stress which resulted in the fracture described.

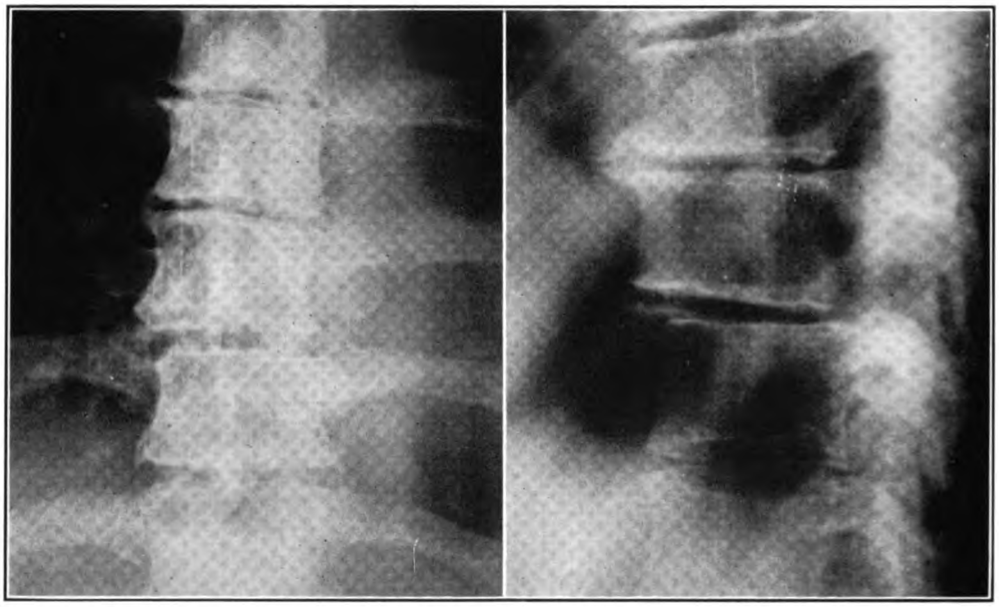

FIG. 251.—Fracture dislocation of the tenth on the eleventh thoracic vertebra, with lateral and anterior displacement of the tenth.

A considerably more frequent group of fractures of the dorsal vertebræ occurs in patients undergoing shock treatment for psychiatric disturbances. Forceful anterior flexion during the convulsive episode may produce compression fractures of the anterior aspects of one or several upper thoracic vertebræ, usually the fourth, fifth or sixth. No changes of the intervertebral discs are associated with these compressions. Alterations in vertebral contour usually are more readily identified on the lateral than the frontal views.

However, compression of the right or left sides can only be seen on frontal roentgenograms. Such fractures were reported by Vogl and Osborne (1940) in vertebral compression fractures sustained during convulsions following metrazol therapy. Similar changes were reported by Krause and Langsam (1941), both indicating that the fractures were more common in the upper dorsal spine. The latter authors reported an incidence of 42.6 per cent out of 75 consecutive cases. Fractures following electric shock therapy were reported by Lingley and Robbins (1947) in 23 per cent of 230 cases. Meschan, Scruggs and Calhoun (1950) reported an incidence of vertebral injury of the thoracic spine in 35.4 per cent of their cases, with an average of 2.6 vertebræ involved per patient. The third, fourth and fifth dorsal vertebræ appeared to be affected more frequently than any of the others. These authors noted that a high correlation existed between anterior narrowness of any vertebral body on the control films and the subsequent occurrence of vertebral fracture, although very few of the narrowed vertebræ themselves were fractured. They observed that osteochondrosis seemed to provide some resistance against fracture, the incidence of fractures under such circumstances being low. Funkhouser and Davis (1952), in a review of 50 spinal fractures due to electric shock treatments, noted an incidence of approximately 20 per cent, and that men were affected more frequently. The third, fourth and

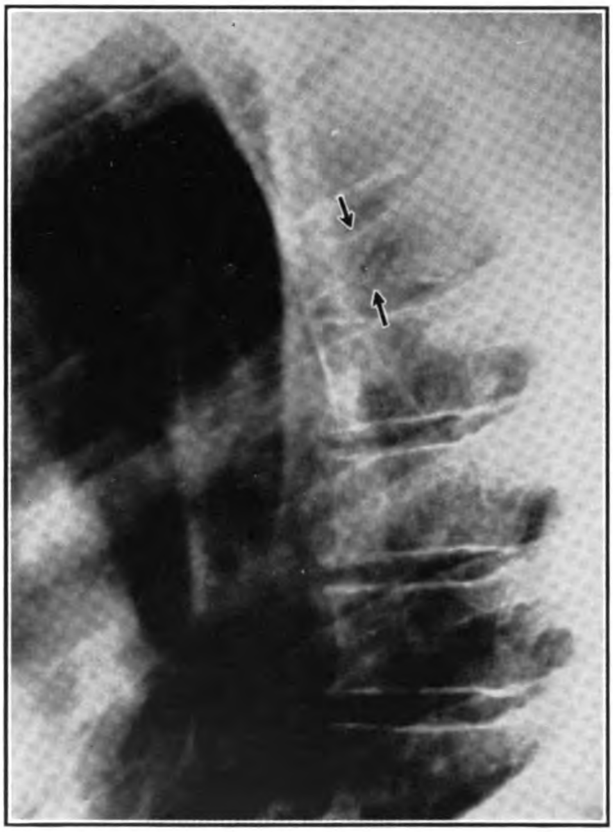

FIG. 252.—Postconvulsive fracture of the body of the fifth thoracic vertebra (arrows).

fifth thoracic vertebræ were involved most often. Pain with these fractures, incidentally, is absent or mild.

It has long been known that major epileptic convulsions may cause fractures of the dorsal spine by acute hyperflexion. In 1938 Rand reported on post-tetanic gibbus in 3 children with varying degrees of collapse of the bodies of the midthoracic vertebræ. Radiographic examination six months later showed kyphosis with marked narrowing of the fifth, sixth and seventh thoracic vertebræ in a twelve-year-old girl who recovered after anti-tetanus serum, and similar observations were made in 2 other children.

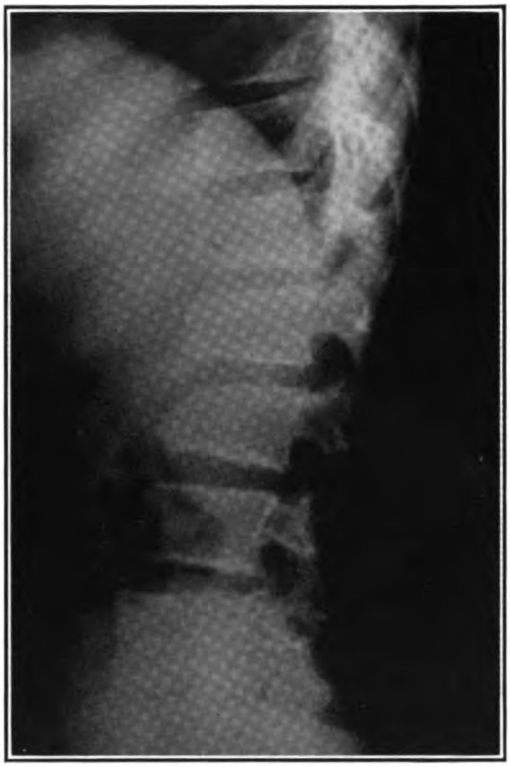

FIG. 253.—Old healed fracture of anterior aspect of the first lumbar vertebra incurred during rachitic tetanic convulsion during infancy.

Dietrich, Karshner and Stewart (1940) reported on 28 cases of tetanus with characteristic compression fractures of the midthoracic vertebræ. They noted the wedge-shaped appearance of the involved vertebræ, and mentioned that the intervertebral spaces were unchanged. Zürcher (1948) also noted that the intervertebral discs remained unaltered following post-convulsive fractures.

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FRACTURES OF THE LUMBAR SPINE

Inasmuch as fractures of the upper lumbar spine are frequently accompanied by fractures of the eleventh and twelfth thoracic vertebræ, the present section will include mention of injuries to the thoraco-lumbar junction. The mechanism for such injuries is the same as that for fracture of the cervical or thoracic spine, namely, hyperflexion injuries incurred during various types of accidents. While only a single vertebra may be involved, it is more common to observe compressions of two or sometimes

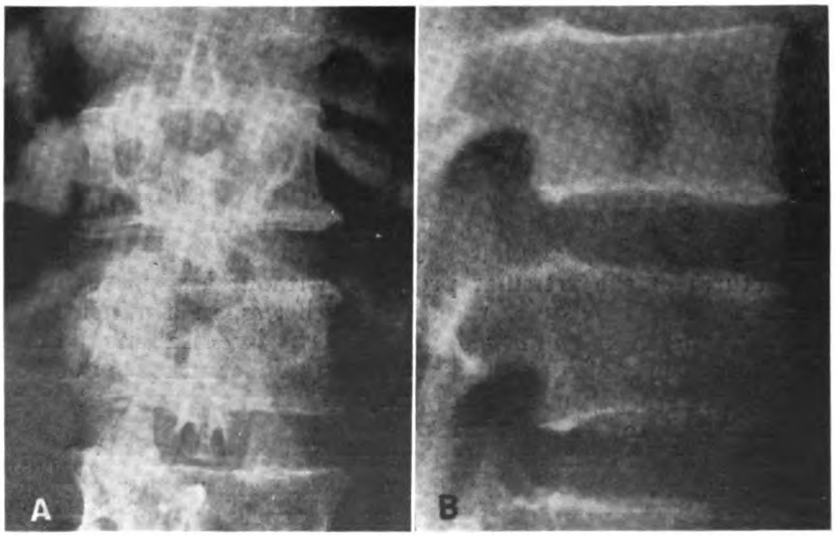

Fig. 254.—Compression fracture involving the right lateral and anterior aspects of the body of the third lumbar vertebra. The vertebral body is wedged anteriorly. There is no change in the height of the posterior aspect of the body as seen on the lateral projection.

three segments, particularly if the thoracolumbar junction is involved. Concomitant injury to the ligaments, capsular structure, soft tissues and intervertebral discs are frequent, but symptoms depend on the effects of local hemorrhage, laceration and cauda equina and nerve root distortions.

There is a relatively higher incidence of paraplegia associated with injuries to the thoracolumbar junction than observed in the lower lumbar region, ascribed to the difference in the configuration of the spinal cord at the thoracolumbar level. The extent of skeletal damage is not indicative of the degree of cord trauma.

Fractures of the lumbar vertebræ usually are of the compression variety, with narrowing of the anterior aspect of the vertebral bodies. Depending upon the degree of torsional force, fractures of the lateral aspects of the bodies also occur. When a considerable degree of rotational injury is encountered, fractures may also involve the neural arches. In such cases one must recall that anomalous fissures at the distal tips of the inferior articular processes may simulate such fractures. According to Bailey (1937) fractures of the articular processes without associated injuries of the vertebral bodies are quite rare. Nevertheless, in a patient who has suffered a recent injury followed immediately by disabling pain, small comminuted fractures of the neural arches may be identified. Body section and oblique stereroentgenographic study of the lumbar spine is almost essential in identifying these injuries.

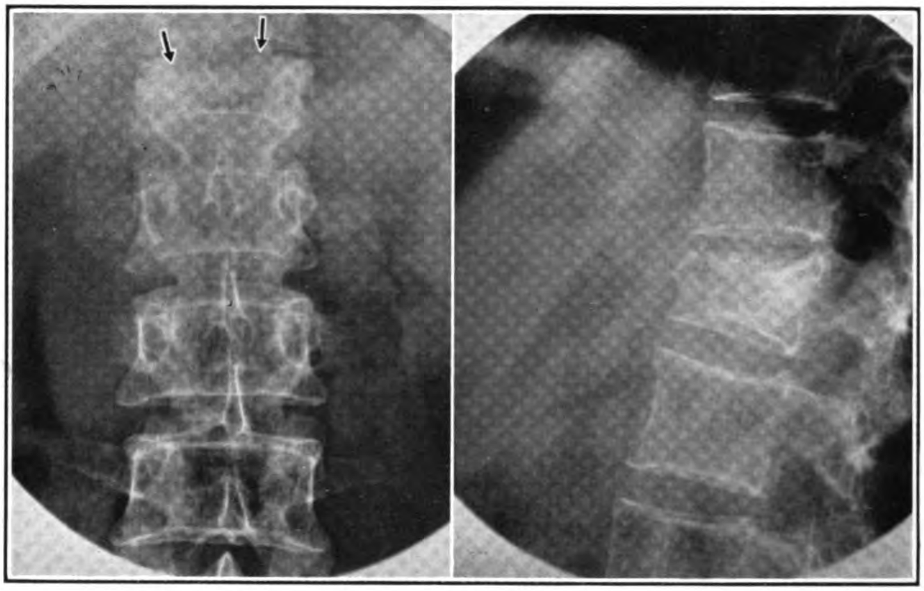

FIG. 255.—Impacted fracture through the superior surface of the body of the first lumbar vertebra (arrows).

Occasionally one encounters what appears like small avulsion fractures of the anterior aspect of a vertebral body. These are usually accompanied by narrowing of the anterior or anterolateral aspect of that vertebral segment. One must regard with suspicion these small irregular triangular shadows seen in the anterosuperior aspect of normal vertebræ. In such instances, even though there is a history of recent trauma, the change is one

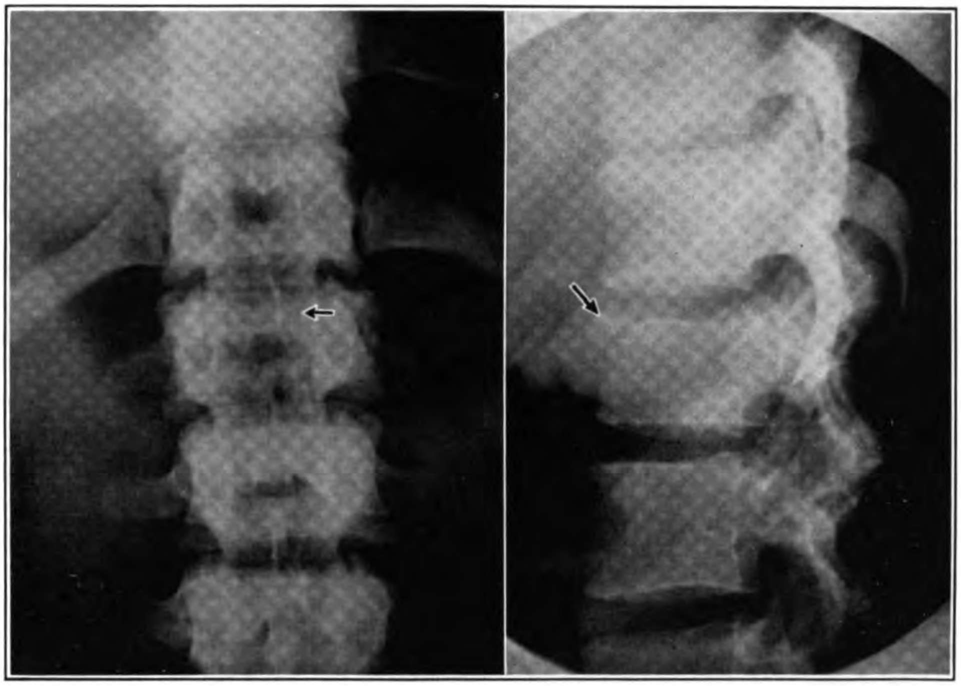

FIG. 256.—Fractures through the left lamina of the twelfth thoracic vertebra (arrows) and the anterosuperior aspect of the body of the first lumbar vertebra.

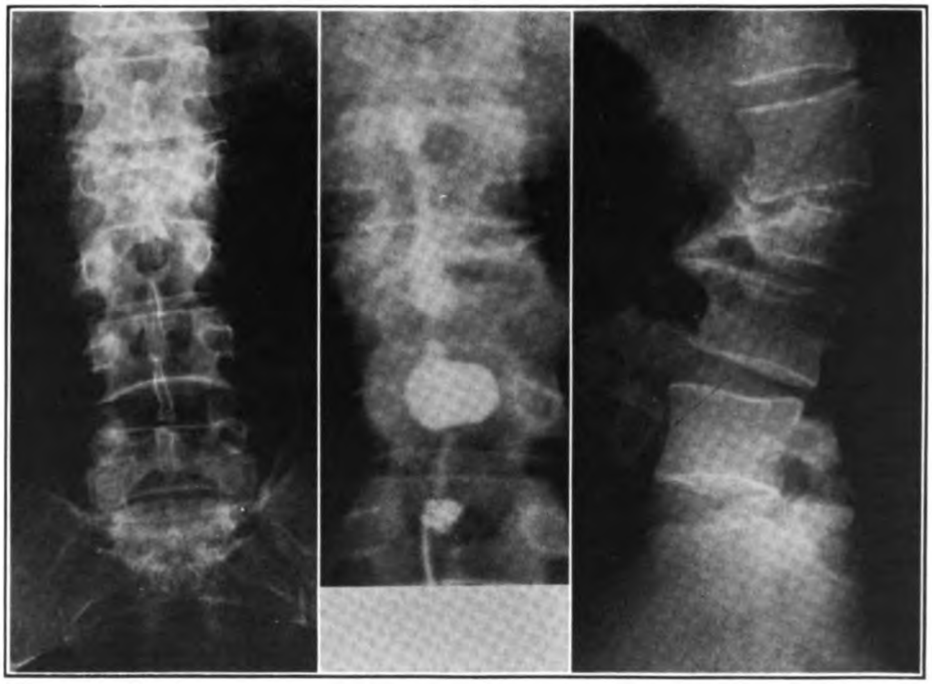

FIG. 257.—Old compression fracture of the anterior aspect of the second lumbar vertebra, with myelographic block and heavy bony bridging between the first, second and third lumbar vertebræ.

associated with a disturbance of the intervertebral disc rather than a fracture, and is of no clinical importance.

In distinguishing between old healed compression fractures and healed nontraumatic lesions of the lumbar spine, Ellis (1944) mentioned that with fractures the wedge deformity is roughly preserved. This affects the upper half of the vertebral body alone, or the upper half out of proportion to the lower half of the body. Increased density of the spongiosa may persist for a long time in these patients. He believed that osteophyte production and generalized spondylosis did not result from compression fractures of a vertebra. Baab and Howorth (1951), however, mentioned that a majority of patients with mild compression fractures in the thoracolumbar region, and all with a severe type of fracture, eventually had spontaneous anterior bony ankylosis. They also noted that patients with mild or severe fractures presented posttraumatic thinning of the intervertebral discs. Osteophyte formation at the fracture sites could not be correlated with symptoms. Patients with spur formations prior to fracture had their symptoms somewhat longer, but the end results were the same as with other patients.

Fractures of the transverse processes of the lumbar spine are readily identified when there is displacement of fracture fragments. However, in many cases the fracture line are difficult to identify because of overlying soft tissue and gas shadows. In such instances the patient should be reprepared and the study repeated, preferably with the aid of a compression band to help eliminate overlying gas shadows. Spinous process fractures of the lumbar spine are infrequent, but should be considered whenever a point of tenderness is found over one of these structures. It is helpful to view all lateral lumbar spine films through a bright light to check on the spinous processes, inasmuch as these are usually obscured because of relative overpenetration. Special cone-down views with a lighter technic may be of further assistance.

A case in which pseudarthrosis followed fractures of the transverse processes of the second to fifth lumbar vertebræ was reported by Hyman (1945). This patient had pain three months after having been immobilized for this injury. Re-examination showed joint formations at the fracture sites. At operation these joints were enclosed in a capsule, and section through the pseudarthrosis showed a fibrous band between the bony masses which prevented union. A comparatively well-organized hyalin cartilage covered the joint surfaces.

Subluxations of the apophyseal articulations of the lumbar spine are considered a cause of pain because of the production of tension on the capsular ligaments, encroachment of the articular facets on the lumen of the intervertebral foramina, and because of erosion and sclerosis of the apposing joint surfaces (Hadley, 1935).

The advantages of pantopaque myelography in investigating penetrating wounds of the spinal canal were discussed by Hinkel and Nichols (1946). They advocated this procedure because of information made available concerning the location and actual nature of an injury. Only a small number of patients suffering from penetrating wounds have actual perforation of the dura and extensive cord damage. More often the injury is in the nature of a hemorrhage, dislodged bone chips and infection. All of these may be remedied and present a better prognosis than does true destruction of the cord or cauda equina. Included among myelographic defects observed after spinal trauma are sharply localized, clearly marginated indentations, angulations of the dural sac with displacement, and feathery irregular filling defects which may be part of the picture of an arachnoiditis. Complete spinal canal block may also be observed. The passage of pantopaque outside of the spinal canal which might be interpreted as due to faulty technic may be due to a tear in the arachnoid. In their cases the escaping contrast medium never flowed freely into any other space, but outlined a blind pocket. This they described as an extra-arachnoid puddle, with its long axis parallel to that of the neural canal. When myelography disclosed a diverticulum-like contour, the possibility of a local collection of fluid causing compression of the spinal canal or the adjacent nerves had to be considered. Avulsion of nerve roots produces a similar change.

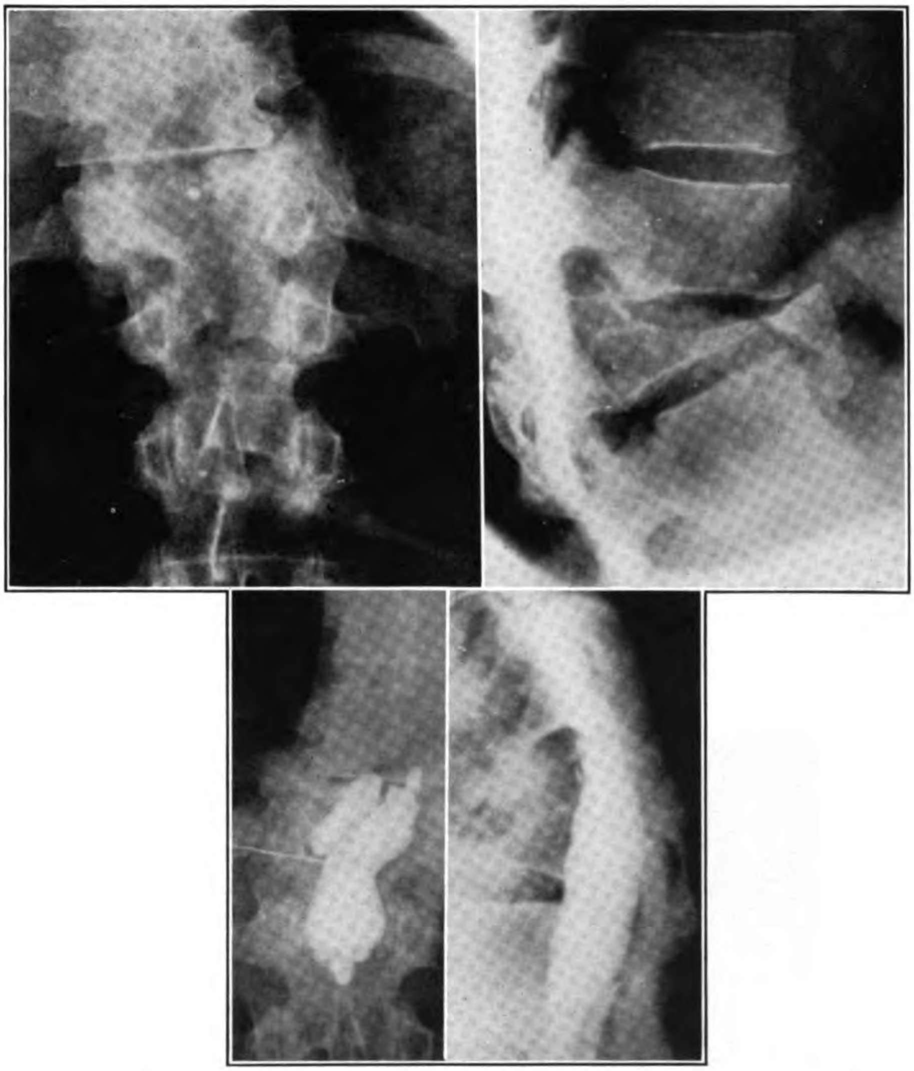

FIG. 258.—Old compression fracture of twelfth thoracic vertebra with paraplegia. A mild gibbus and myelographic block were present. (Courtesy of Dr. Ralph Goldin, Albert Einstein College of Medicine, New York City.)

We do not hesitate to use myelography in cases of compression fractures associated with injuries to the neural arches when neurologic symptoms are present. In such cases defects due to protrusion of bony fragments have been noted and prompt surgical intervention resulted in relief of symptoms.

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FRACTURES OF THE SACRUM AND COCCYX

These usually follow direct trauma, most often secondary to either a fall or to a direct blow. Characteristically, these patients complain of a considerable degree of pain which is exaggerated by attempting to walk or stand, and becomes increasingly acute when the patient places weight on the injured area. Radiographic examination is often difficult to evaluate. We have found it advantageous to make several anteroposterior studies in various angles of projection in order to bring out the various sacral and coccygeal segments. The fracture lines may be difficult to identify because of overlying gas and fecal shadows, even after adequate preparation of the patient. The lateral views have proved to be of more help in diagnosing

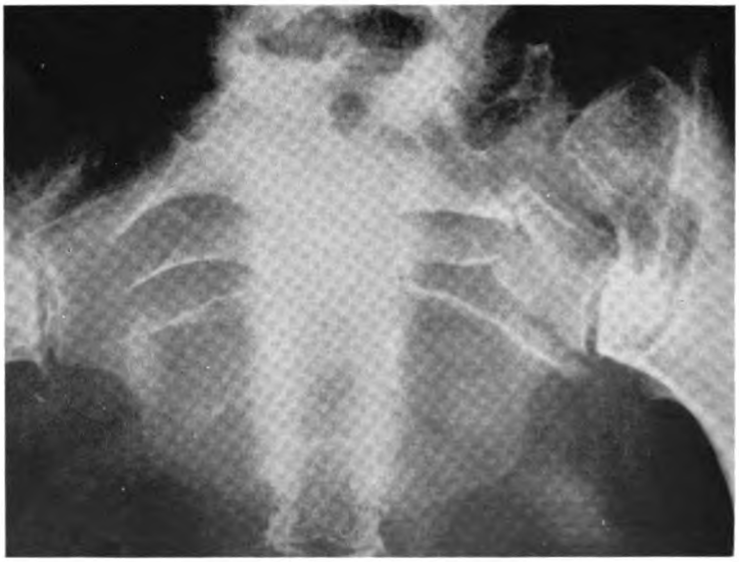

FIG. 259.—Fracture through the left sacral wing passing through the superior first and second foraminal margins and the left side of the sacrum just beneath the sacroiliac joint.

such fractures. Tears of the sacrospinous and sacrotuberous ligaments may be diagnosed when there is avulsion of their insertions into the lower sacrum (Burman, 1952).

In addition to the bony injuries incurred in fractures of the sacrum there may be neurologic complications because of trauma to the cauda equina. Bonnin (1945) in a review of 44 pelvic fractures, noted that the sacrum was involved in 45 per cent. He mentioned that the weakest portion of the sacrum was at the first and second foramina, where in his experience the fracture lines usually were found. He also observed that the fractures sometimes were not visible until several weeks after the injury. The neurologic manifestations described by Bonnin included evidences of damage to the first and second sacral nerves, as evidenced by weakness of plantar flexion of the ankle, loss of ankle jerk reflexes and loss of the power of the hamstrings and glutei muscles. Marked paralysis of the biceps femori is frequent, and the calf and hamstring muscles may be tender. Altered sensations and referred pain to the outer side of the calf and foot may be present.

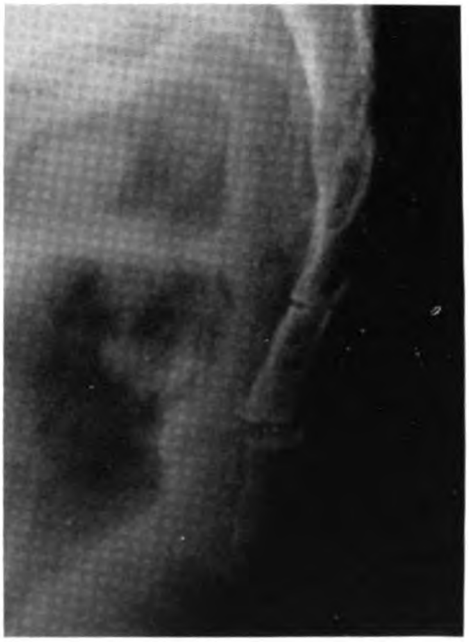

FIG. 260.—Posterior dislocation of the first coccygeal segment following a fall.

In our own experience fractures of the sacrum were somewhat more frequent in the third and fourth sacral segments. Neurologic deficits were infrequent. The usual complaint was pain, often unrelieved when lying down, and aggravated by motion. Sharp pain was often elicited on rectal examination.

An unusual complication of a sacral fracture was described by Dobrzaniecki and Haak (1942) in a twenty-six-year-old man who fell from a tree, striking the ground in a sitting posture. A fracture of the right side of the sacrum was found. Intrathecal injection of lipiodol showed globules along the line of fracture, gathering within a protruberance descending into the intragluteal furrow. Rectal examination revealed an elastic projection of the posterior wall, and the finger could palpate along the fracture line. This was considered to be a spurious meningocele following trauma. The mass-like lesion disappered under conservative treatment, and no operation was deemed necessary.

A fracture dislocation of the sacroiliac joint associated with rupture of the symphysis pubis, presumably produced by the force of uterine contraction during labor was reported by Urist (1953). In this patient an associated injury to the first sacral nerve root was produced. The plain roentgenograms showed the fracture line in the superior aspect of the left side of the sacrum adjacent to the sacroiliac joint. Myelography showed a displacement of the caudal sac contralaterally.

Fractures and fracture dislocations of the sacro-coccygeal area are quite frequent. They usually follow trauma, and are painful out of proportion to the degree of osseous injury. The fracture lines sometimes are difficult to identify, and may become more apparent on roentgenograms made some time after the accident. The pain may be persistent and require surgical relief.

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TRAUMATIC AVULSION OF NERVE ROOTS

Murphy, Hartung and Kirklin (1947) reported 3 cases in which myelographic examination of the cervical spine revealed lateral collections of pantopaque. These were not lateral extensions of the nerve sheaths, but rather pantopaque extending into an area created by rupture of the meninges which permitted escape of the opaque medium into the adjacent tissues. These authors suggested the term "traumatic meningocele" even though they had no way of stating that the outpouchings actually were lined by the meninges. No operative confirmation was made in these 3 cases. In 1950, Whiteleather and Clayton reported 3 additional cases, 1 of which was confirmed at operation. This patient, a fourteen-year-old boy, had been struck on the left shoulder and upper arm in a downward direction by a whirling airplane propeller, resulting in considerable injury to the soft tissues, fracture of the left clavicle and humerus, and loss of motor and sensory functions in the area innervated by the left brachial plexus. On myelographic examination an oval collection of pantopaque was found in the lateral aspect of the interspace between the fifth and sixth cervical vertebræ, the collection being separated from the main body of the pantopaque column within the cervical canal by a hiatus of about 1 cm. A preoperative diagnosis was made of a traumatic avulsion of the left seventh and eighth cervical and first thoracic nerve roots with formation of a traumatic meningocele at the level of the sixth left interspace. Exploration of the left brachial plexus revealed that the sixth and seventh cervical nerve roots were surrounded by a hard, dense scar. The eighth root had been pulled through the seventh intervertebral foramen and the torn end was found 1 cm. lateral to the foramen. In the other 2 cases, lateral collections

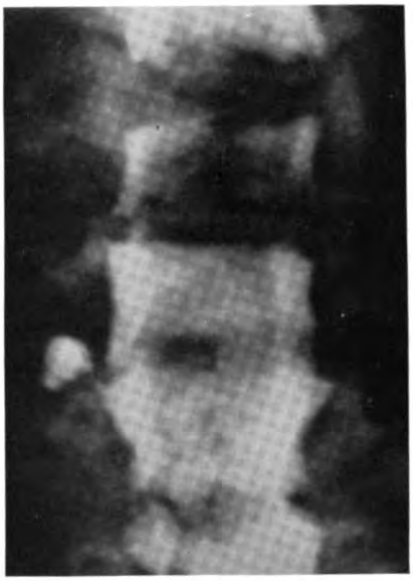

FIG. 261.—Avulsion of the nerve root of the right sixth cervical nerve. Note the pooling of pantopaque lateral to the column.

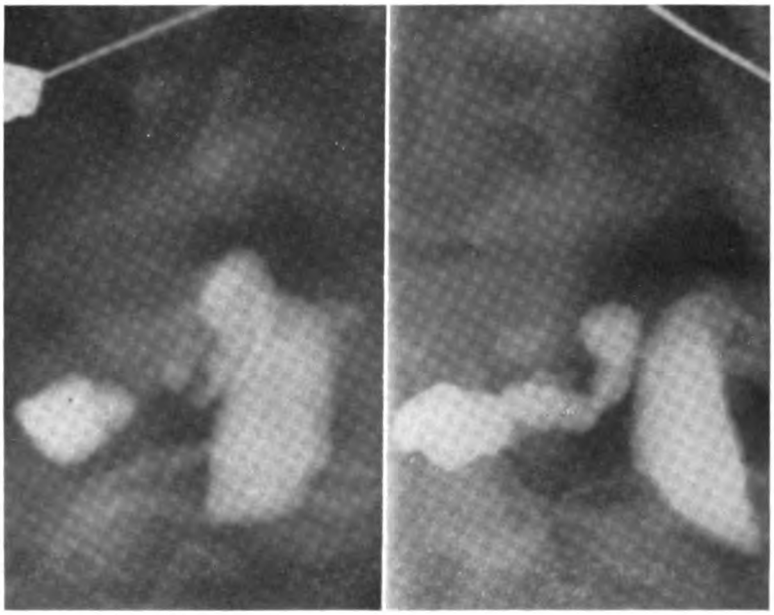

FIG. 262.—Traumatic avulsion of fourth lumbar nerve root. The pantopaque has collected in an irregular cavity on the lateral aspect of the column, and can flow freely back into the subarachnoid space. (From Finney, L. A. and Wulfman, W. A., Am. J. Roentgenol., **84**, 952, 1960, courtesy of Charles C Thomas, Publisher.)

of pantopaque were seen outside of the spinal canal, extending into the intervertebral foramen. It was deemed inadvisable to operate on the latter 2 patients.

The subarachnoid space in the cervical region does not extend beyond the axillary pouch. As seen on myelograms the pouch is visualized but the root sleeve is not. If there is a tear of the axillary pouch the pantopaque passes through the intervertebral foramen into a diverticulum-like cavity. With avulsion of a nerve root, that structure or its rootlets are torn from the spinal cord. Separation of a nerve root implies rupture of its continuity. Inasmuch as the nerve roots are more fragile than their investing membranes, situations can occur in which the roots are torn but the investing membrane remains intact. In such cases myelographic examinations are likely to be normal (Jaeger and Whitley, 1953).

Traumatic avulsions of the lumbar nerve roots are rare. Finney and Wulfman (1960) reported such a case secondary to a flexion-abduction injury to the hip with concomitant traction damage to the common peroneal nerve and the intradural fourth lumbar nerve roots. The myelographic picture was comparable to that seen with brachial plexus avulsions.

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POSTLAMINECTOMY DEFORMITIES

It has been noted that following bilateral laminectomies changes appear in the intervertebral discs and the associated vertebræ. Taylor (1927) observed that following bilateral laminectomy of the cervical spine, deformities of the intervertebral discs and adjacent vertebral bodies occurred, associated with weakness and pain. Horowitz (1941) reported obvious deformities following laminectomy of from 2 to 5 vertebræ in which the spinous processes and laminæ on both sides had been removed, but in which the articular facets had been preserved. These observations, over a period of from eight months to fifteen years, revealed changes in the intervertebral discs and vertebral bodies in the region of the lumbar laminectomies. These were narrowing of the discs, kyphosis and sometimes bony proliferations at the margins of the vertebral bodies.

In 1 of our cases in which a cervical laminectomy had been done for relief of an Arnold-Chiari malformation, the patient developed severe pain after operation. Radiographic examinations over a period of one year showed progressive decalcification and irregularity of the bodies of the second to fifth cervical vertebræ. The usual lordotic curve vanished, the intervertebral spaces became irregularly narrowed, and the margins of the involved vertebræ became irregular. However, with orthopedic treatment the cervical curve straightened and the vertebral bodies regained their osseous densities. Spur formations between the bodies resulted in a stable cervical spine after about three years.

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KÜMMEL'S DISEASE

This condition refers to posttraumatic compression of a vertebral body either in the thoracic or lumbar region, which usually is unidentifiable immediately after the injury. The degree of collapse is usually mild, and becomes apparent only several months after the injury (Kümmel, 1895, 1928). The process is believed to be secondary to decalcification of bone because of interference with the blood supply to the vertebra. According to Schinz and his co-workers (1952) this lesion is of practical importance. They mentioned that histologic examination of the affected vertebræ may reveal infractions of the cartilaginous laminæ, hemorrhages into the medullary tissues and microfractures and necrosis of the trabeculæ. The intervertebral discs may also be fragmented, with protrusions of cartilaginous material into the cancellous tissue. They also mentioned that the intervertebral disc eventually may undergo calcification, simulating synostosis of the adjacent vertebral bodies.

There appears to be some difference of opinion as to whether Kümmel's disease actually represents a true clinical entity. Morton (1943) stated that there was still doubt as to its existence, and cautioned against the diagnosis unless one were very sure of the facts involved in the history. He stressed the sequence of injury, normal roentgenographic examination of the spine followed by slow, progressive wedging of an involved vertebra associated with pain.

Fletcher (1947) pointed out that wedging of vertebral bodies occurs as a normal variation, and believed that epiphysitis is probably a more common cause of plain wedging than compression fractures. While he made no direct reference to Kümmel's disease, his statistical study of uncomplicated wedging in a group of 550 adult male veterans under fifty years of age is of interest when a diagnosis of Kümmel's disease is considered.

In our own experience we have frequently encountered wedging of the thoracic or upper lumbar vertebræ in individuals referred for examination of the spine for reasons other than trauma. In such cases we mention that the wedging is present, and further state that unless the history is absolutely characteristic the process is most problably the result of a disturbance of the intervertebral discs such as Scheuermann's disease.

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PARAPLEGIC NEUROARTHROPATHY

Following injuries to the spine with consequent paraplegia, neuropathic articular and periarticular changes may occur in the lower extremities. As pointed out by Solovay and Solovay (1949), these include intraarticular destructive and productive changes, and also extra-articular soft tissue ossification. They described changes in the lower extremities considered representative of Charcot's neuroarthropathy. The mechanism they postulated was a long continued pressure over an anesthetic bony prominence and lack of warning sensations, which they believed to be equivalent to trauma in initiating ossification in periarticular tissues.

Abramson and Kamberg (1949) noted similar changes in the soft tissues, chiefly about the hips, and erosive bone changes around the trochanters. They also pointed out an additional change, a spondylitis which appeared to be in many ways like rheumatoid arthritis, with frequent involvement of the diarthrodial joints. They noted that lesions below the first lumbar vertebra were not associated with such changes. With lesions above the first lumbar level the apophyseal joints showed varying degrees of fusion, some with osteoporosis and some with osteosclerosis of the vertebral bodies, others with a combination of the two forms of bony disturbance. They believed that the cause of this was a hyperemia of bone followed by decalcification, diminished circulation, and that the osteosclerosis followed loss of blood supply due to necrosis of bone.

Subjective complaints in these patients were not striking because of the basic lesion of the cord. Heilbrun and Kuhn (1947) pointed out that erosive lesions developed where the soft tissues over bony prominences such as the trochanters became devitalized. Pathologically, they noted that subacute and chronic infection of the tissues appeared overlying the bone, with extension to the superficial layers of bone. This leads to resorption of the cortex and the underlying trabeculæ, and the bone marrow becomes replaced by fibrovascular tissue which accompanies a low grade inflammatory process. They recognized that the neurogenic influences were probably of major importance, even though soft tissue trauma may play an important role in the appearance of these changes.

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SPONDYLOLISTHESIS

By definition spondylolisthesis indicates displacement of one vertebral body on another, usually with disruption of the neural arch of the involved vertebra at the pars interarticularis. This occurs most often at the fourth and fifth lumbar articulations, less frequently higher in the lumbar spine. Meyerding (1938) noted that the displacement was at the lumbosacral interspace in 82.1 per cent of 583 cases. In 11.3 per cent the fourth lumbar vertebra was displaced. Spondylolisthesis of the third lumbar vertebra occurred in 0.5 per cent, and displacement of the second lumbar vertebra was found in 0.3 per cent. He also observed two in which the fifth lumbar vertebra was displaced on a sixth lumbar vertebra (0.3 per cent) and another in which the sixth lumbar segment was displaced anteriorly on the first sacral vertebra. In five cases (0.9 per cent) there was a double spondylolisthesis with displacement of both the fourth and fifth lumbar vertebræ. Reverse spondylolisthesis, with posterior displacement of the involved vertebra, was observed in 26 cases (4.4 per cent), usually at the lumbosacral interspace. Spondylolisthesis occurring in the cervical spine will be considered as a separate entity.

In a rather limited number of cases spondylolisthesis occurs without a defect in the neural arch. This occurs more often in patients with exaggerated lumbar lordotic curves together with malacic diseases, osteoporosis and spondylosis. Changes in the inclination of the articular facets permits a forward displacement of one vertebra on another.

The origin of the defect in the pars interarticularis with spondylolisthesis has caused much speculation. It appears that present evidence favors a traumatic origin rather than a failure of fusion of two centers of ossification. Bowman and Goin (1926) believed that infantile or birth trauma with actual fractures or local osteochondrovascular disturbances of the neural arch were the initiating factors. Chandler (1931) mentioned that ossification of the pars interarticularis progressed by the formation of cyst-like spaces within a thin shell of bone. The pars interarticularis, consisting mainly of cartilage and a large lake of blood vessels during the period of early ossification, was considered particularly susceptible to fracture. In an investigation of two hundred fetal spines Batts (1939) found that there were none with separated neural arches, and postulated that if the lesions were congenital such defects might be anticipated in about the same number of fetal spines as seen later in life, approximately 5 per cent. Rowe and Roche (1953) did not find a single example of a defective neural arch in over 500 stillborn and neonatal cadavers. Lerner and Gazine (1946) favored the idea that isthmic defects resulted from trauma in early life, and commented that fracture through cartilage heals by pseudarthrosis rather than be callus formation. Hadley (1945) mentioned that bony callus was frequently seen as a mass projecting into the spinal canal from the lamina, and believed this to be derived from a break in the lamina. Mutch (1956) described the pattern of ossification in the vertebral arches, noting that the fifth ossifies later than the first. He, too, favored the traumatic etiology of clefts in the pars interarticularis.

Clefts in the pars interarticularis, while often associated with spondylolisthesis, occur without displacement of the involved vertebral body. This condition is referred to as spondylolysis or spondyloschysis (Rhodes and Colangelo, 1946). The term prespondylolisthesis has sometimes been applied to the situation. An interesting mechanism for the production of spondylolysis was postulated by Nathan (1959). He found 19 cases in a series of 450 skeletons. In typical instances the lower lumbar vertebræ were involved. The clefts in the isthmus were located between the downward projecting inferior articular processes of the vertebra above and the upward projecting superior articular processes of the vertebra below. Such clefts appeared to be produced by compression of the isthmuses between these articular processes by a sort of pincers mechanism. Preceding bony abnormalities or congenital lesions were not found. He observed skeletons in which the development of the spondylolysis stopped before total separation of the vertebral arch occurred, with marked depressions on the upper and lower surfaces of isthmuses precisely at the site of contact with the articular processes of the adjacent vertebræ. We have observed four instances in which lumbar spine radiologic examinations in children from one to three years of age disclosed somewhat similar waist-like narrowing in the pars interarticularis of the fifth lumbar vertebra.

According to Brailsford the term prespondylolisthesis, as described by Armitage Whitman, also may be applied when the lumbosacral angle approaches ninety degrees. It was considered that such an exaggerated lordotic curve may precede dislocation. Brailsford felt that this term was unfortunately chosen because exaggerated lumbosacral angles occur in a wide variety of conditions unassociated with spondylolisthesis, such as neglected rickets, osteomalacia and osteogenesis imperfecta. The use of the term prespondylolisthesis to refer either to a cleft neural arch or to an exaggerated lumbar lordotic curve is unsuitable, inasmuch as in neither is spondylolisthesis a necessary concomitant. An anatomic reference appears a more satisfactory way of describing such changes, and avoids the unfounded supposition that another pathologic entity may in time be superimposed.

The anatomic changes seen with spondylolisthesis at operation include looseness of the lamina, permitting it to be rocked back and forth to a variable degree. Some, with lesser mobility, are bound down by fibrous ankyloses, with ill defined masses of fibrous tissue merging imperceptibly with the adjacent ligaments. Mucoid degeneration occurs at these sites, and the bulging of soft tissue towards the intervertebral foramina and spinal canal may be considerable. The foramina become distorted, and the nerve roots in proximity to the displaced inferomedial wall of the superior pedicle may be bound down by adhesions. A step formation forms as the vertebra is displaced. The disc may be considerably narrowed. The dura makes a double bend as it follows the angulated spinal canal, and becomes adherent to the lamina and ligamenta flava. The cauda equina may be stretched quite tautly across the step. When this is accompanied by a narrow lumbar canal and distorted lateral recess of the spinal foramen, small discal herniations or spondylotic spurs may produce disproportionate symptoms.

The incidence of spondylolisthesis has been variously reported as between 3.5 per cent (George, 1939) and 5 per cent (Bailey, 1947). Bailey mentioned that among 2080 lateral lumbosacral roentgenograms in unselected cases, 4.4 per cent showed isthmic defects. Only 0.5 per cent of these complained of low back pain. He concluded that it was incorrect to assume that spondylolisthesis or its presumed precursors in any way were connected with recent trauma. Garland and Thomas (1946) mentioned that in army and navy personnel spondylolisthesis occurred in as many as 10 per cent of cases of low back pain referred for roentgen examination.

Spondylolisthesis has also been reported with far less frequent congenital defects of the neural arch. Included among these by Roche and Bryan (1946) are congenital lengthening of the pars interarticularis, congenital shortening of this region and defects involving one side only of the neural arch. The mechanism for dislocation of a vertebral body in the presence of a unilateral defect in continuity of the neural arch was explained on the basis of a congenitally elongated pars interarticularis on both sides. Brocher (1950) mentioned that spondylolisthesis also occurs because of abnormalities of the articular facets. Heredity was considered one of the causes of spondylolisthesis. Bailey (1947) also believed that heredity played a definite role, and added 3 cases of hereditary transmission to the case described by George (1939).

The youngest case of spondylolisthesis was reported by Kleinberg (1934) in a seventeen-month-old infant. No symptoms were present, and it was presumed that the defect was congenital. This patient also had bilateral dislocated hips. It should be mentioned that Kierulf (1951) quoted Hammerbeck as mentioning a case of spondylolisthesis in a 40-cm. long fetus described by Blum. Hitchcock (1940) reported a case of a four-year-old child. Most cases, however, are seen in the age group from adolescence to adult life, the greatest incidence being between twenty and fifty years of age. In Meyerding's series of 876 cases reported in 1943, the average age was 34.7 years. The youngest was fourteen years and the oldest fifty-nine years. I had seen spondylolisthesis in a child 7 years old, and in patients over 65 years of age.

The clinical picture of spondylolisthesis is one of low back and sciatic pain, often associated with trauma. However, it occurs in advanced forms without any clinical manifestations. When the condition is mild, physical examination may disclose nothing characteristic. With an advanced degree of spondylolisthesis a characteristic dimple often is present in the lower lumbar region, due to depression of the spinous process of the fourth or fifth lumbar vertebræ. In Meyerding's (1938) cases, 63.1 per cent of the 583 patients complained of backache, 17 per cent had backache and pains in the legs, 7.7 per cent had hip pain also. Paralysis was observed in 1.9 per cent.

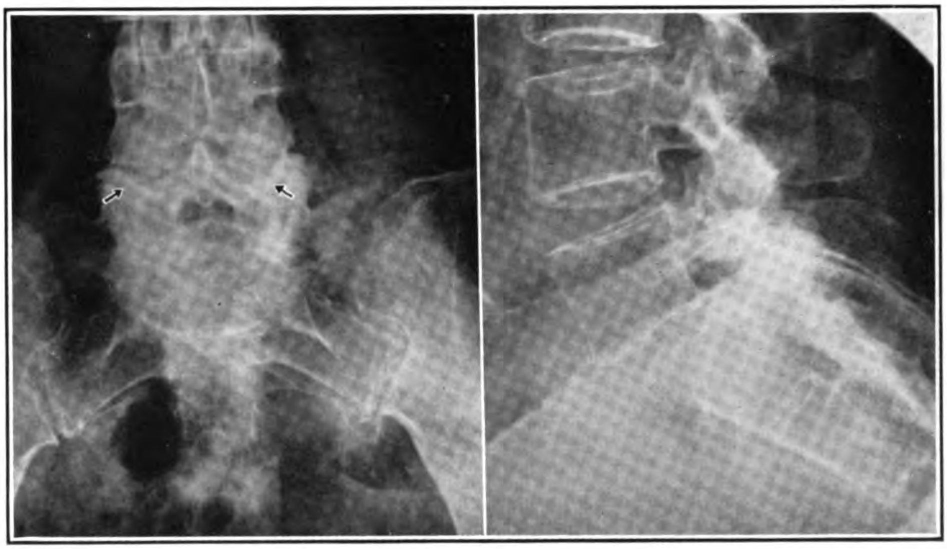

FIG. 263.—Grade I spondylolisthesis of fifth lumbar on the first sacral vertebra. Note the heavy spondylitic reaction at the articular facets bilaterally (arrows).

Meyerding divided spondylolisthesis into four groups. Stage one is considered present when there is only spondylolysis, with no displacement of the vertebral body. Stages 2, 3 and 4 respectively refer to the degree in which the body of the sliding vertebra has been displaced forward. The underlying surface is divided into three parts, and forward slip of up to one third is considered stage 2. Forward slip between one and two thirds is considered stage 3 and from two thirds forward to a complete displacement is regarded as stage 4. Others regard spondylolysis as a separate entity, and divide the degree of forward displacement into four grades, each representing one quarter of the surface of the underlying vertebra.

The question often arises as to whether spondylolisthesis is progressive. Hitchcock (1940) presented 3 patients, four, thirteen and forty-five years of age, in which progressive slipping occurred. In the first case the initial films showed an isthmic defect without displacement. Over an eight-year period this progressed to a frank spondylolisthesis, with marked increase in the interval between the pedicles and the separated laminæ of the affected vertebræ. In the thirteen-year-old patient a grade 3 spondylolisthesis progressed to grade 4 in two years. In the forty-five-year-old woman a grade 1 became a grade 2 spondylolisthesis in three years. In none was trauma significant.

Kierulf (1951) reported a thirty-year-old man who in 1939 had spondylolysis with no displacement. In 1950 a definite spondylolisthesis was present, with decrease in the intervertebral space, narrowing of the intervertebral foramina and forward slipping of the fifth lumbar vertebra.

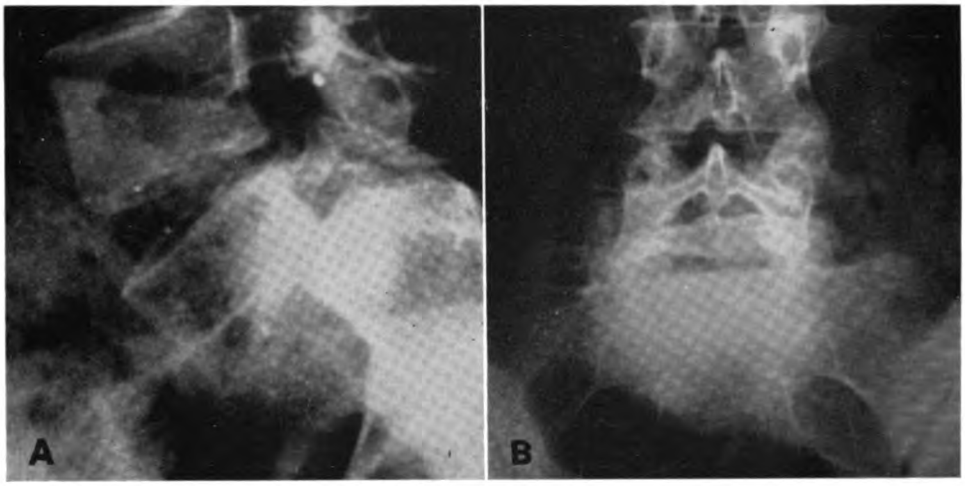

FIG. 264.—Grade III spondylolisthesis of lumbosacral articulation. A heavy sclerotic reaction is present in the posteroinferior aspect of the fifth lumbar vertebra and the anterosuperior articular surface of the sacrum. Note the approximation of the neural arches of the fourth and fifth lumbar vertebræ, and the density of the upper midsacrum (*B*).

We have observed 2 cases in which a posterior shift of the vertebral body occurred. The first was in a fourteen-year-old girl with spondylolysis who developed grade 1 spondylolisthesis associated with low back pain. After being put to rest the fifth lumbar vertebra returned to its normal position. A similar occurrence took place in a twenty-year-old male.

The radiologic investigations of patients suspected of spondylolisthesis should include, in addition to the usual anteroposterior and lateral films, special cone-down views of the lumbar segments involved in the oblique as well as the lateral positions. The oblique views are desirable for portrayal of the articular facets and the pars interarticularis. Lateral roentgenograms often will disclose defects in the pars interarticularis, and a review of our material showed that these defects were seen as often on the lateral as on the oblique films, and sometimes to better advantage.

Frequently associated with spondylolisthesis is a varying degree of narrowing of the intervertebral disc together with sclerotic changes of the apposing vertebral surfaces. In about a third of our cases, particularly in

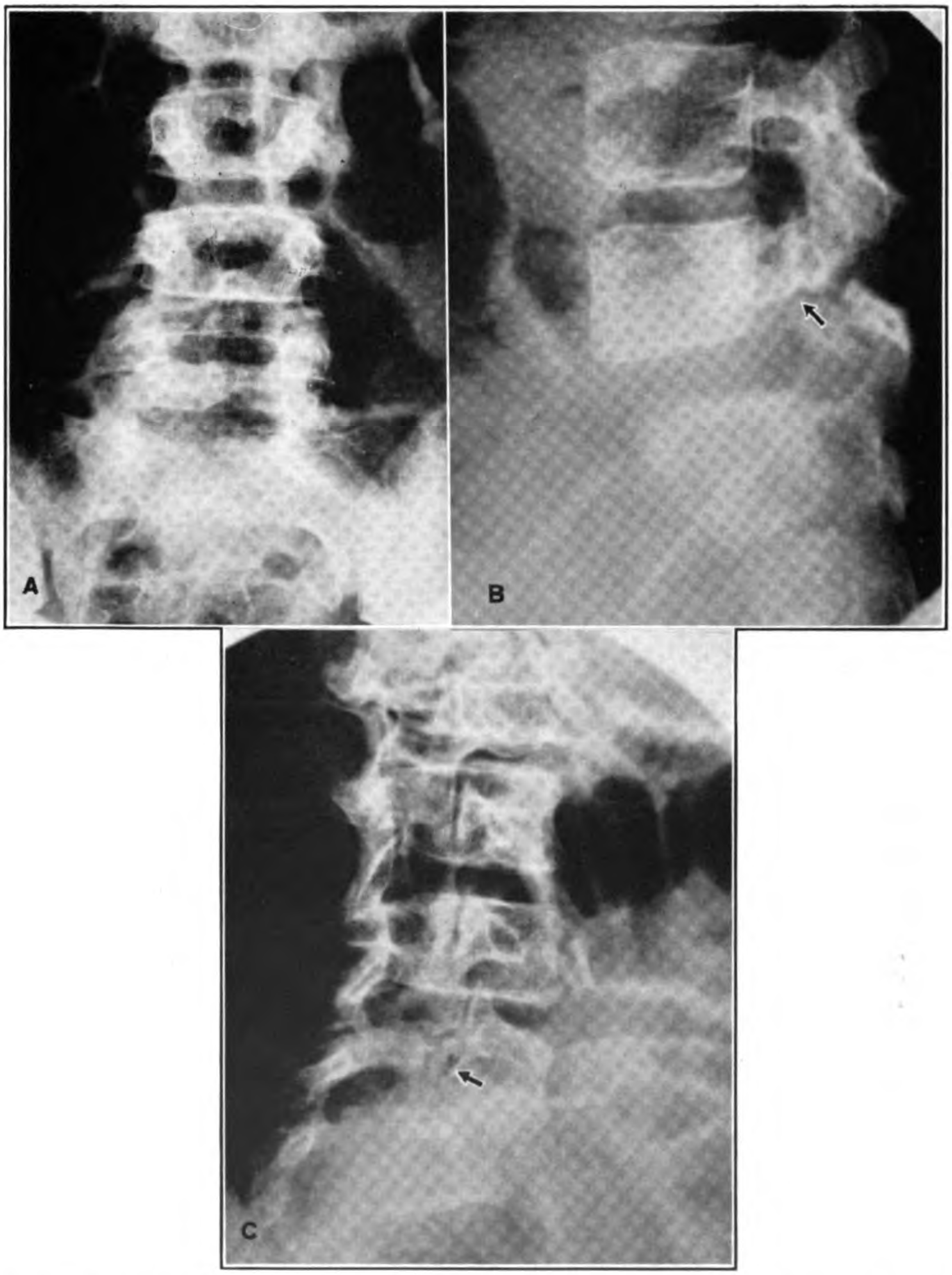

Fig. 265.—*A*, Grade I spondylolisthesis in a twelve-year-old girl. The anteroposterior view is not diagnostic. *B*, Same case, lateral view. A defect is present in the pars interarticularis (arrows). *C*, Same case, oblique view. The articular facets at the lumbosacral joint are inclined forward (arrows). The defect in the pars interarticularis is not as well seen as in Figure 265 *B*.

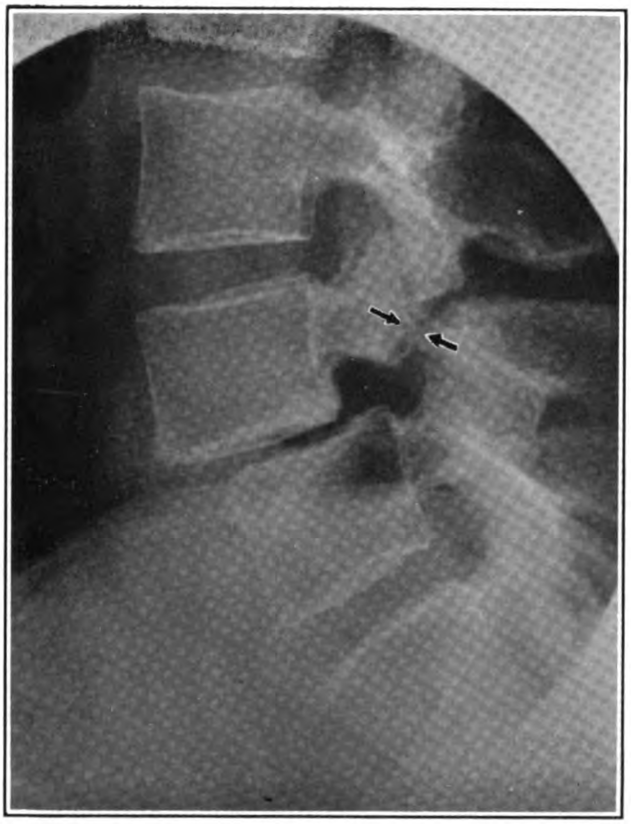

FIG. 266.—Spondylolisthesis of the fourth on the fifth lumbar vertebræ, with marked narrowing of the intervertebral space. The defect in the pars interarticularis is quite large (arrows).

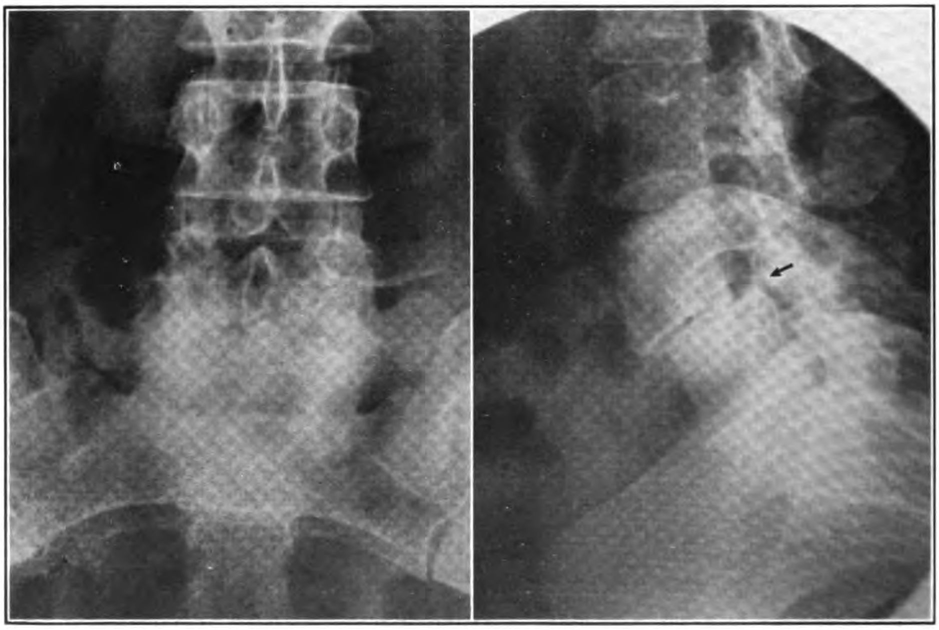

FIG. 267.—Grade II spondylolisthesis of the fourth on the fifth lumbar vertebræ. Considerable hypertrophic change is present at the neural arches, projecting towards the spinal canal (arrows).

patients over thirty years old, hypertrophic changes could be seen. In an equal number of cases sclerotic changes were observed along the articular facets of the lower lumbosacral spine. In some, bony bridging between the anterior aspects of the fifth lumbar and first sacral segments was extensive, and served as a supporting buttress. Such spur formation usually was encountered with grade 3 or 4 spondylolisthesis. In 9 cases with pronounced narrowing of the involved interspace, a concavity developed in the inferior aspect of the fourth or fifth lumbar vertebra into which

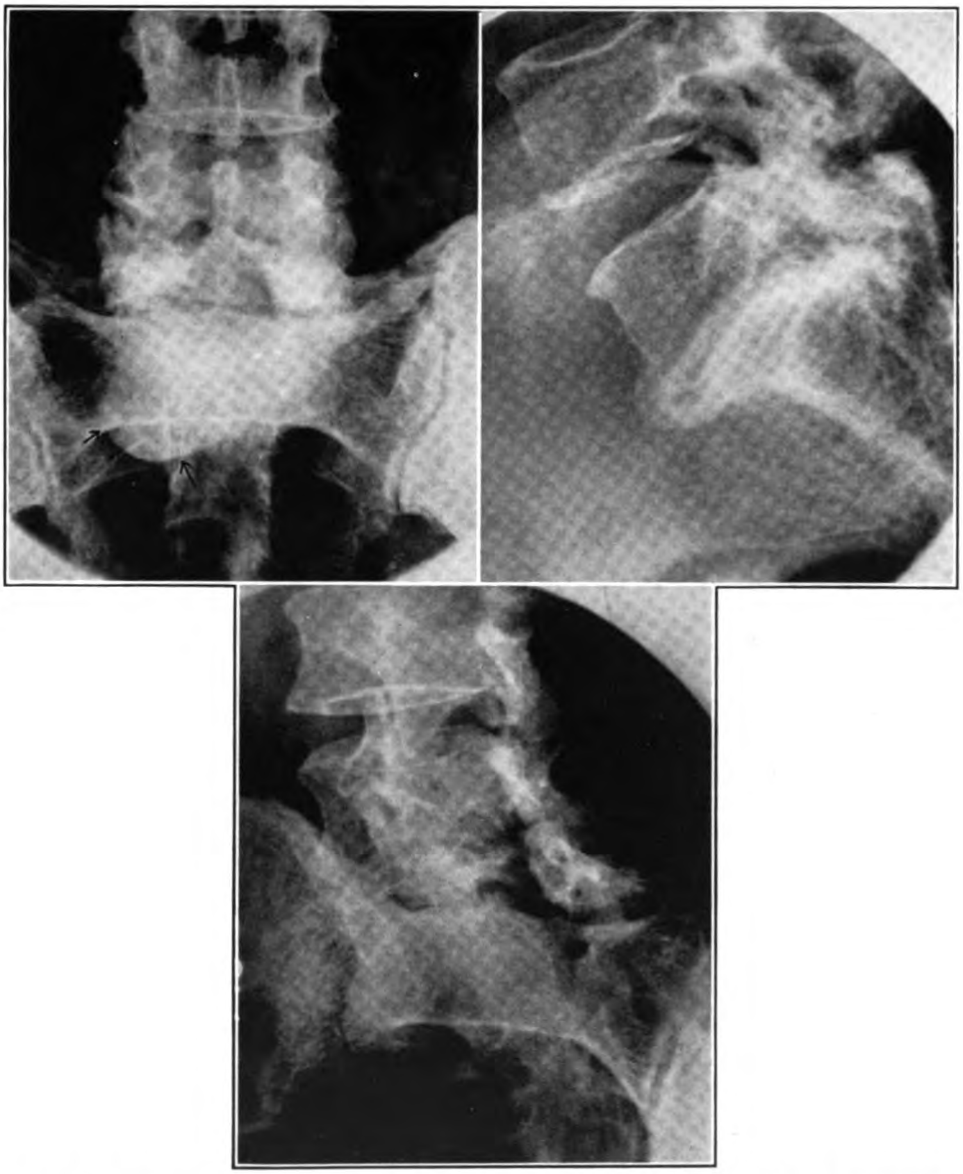

FIG. 268.—Grade II lumbosacral spondylolisthesis with heavy bone bridging on anterolateral aspect of the joint. This produces a bowlike density in the anteroposterior view (arrows). The interspace is markedly narrowed, with sclerotic changes. The bony defect in the pars interarticularis is overshadowed by the bony proliferation.

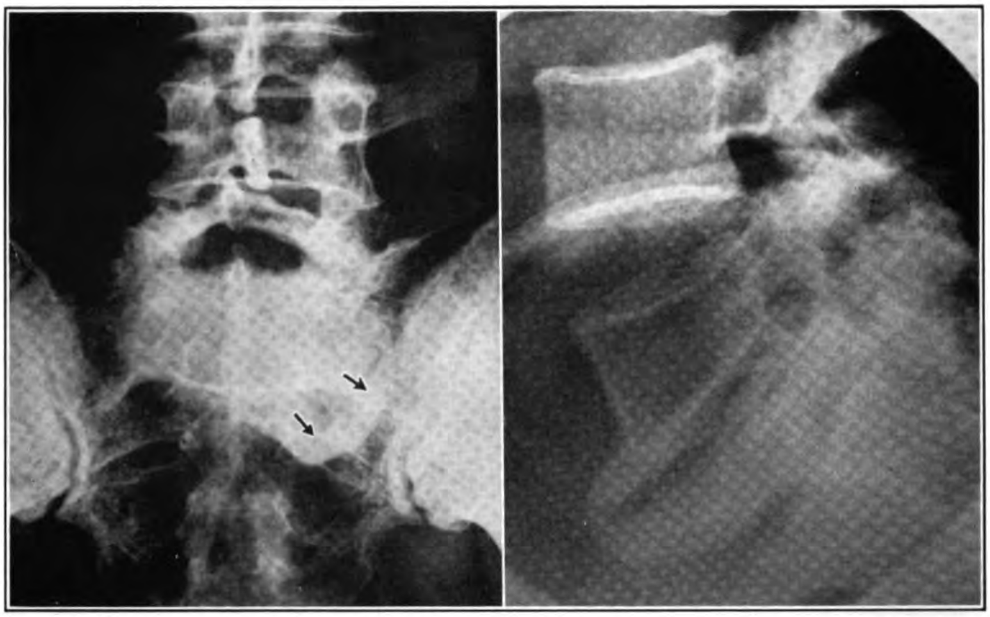

FIG. 269.—Grade III lumbosacral spondylolisthesis with heavy bony bridging on the left anterolateral aspect of the joint (arrows).

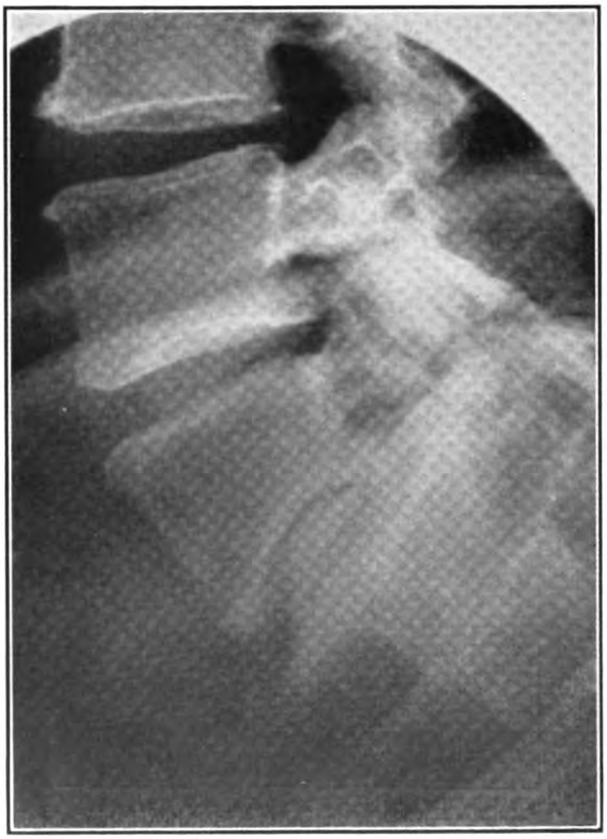

FIG. 270.—Grade III spondylolisthesis with ball and socket arrangement of the articulation.

projected a semicircular prominence from the upper aspect of the underlying segment. This in effect formed a ball and socket joint which blocked further displacement of the vertebra. This corresponds to the term "Torus Promontoralis Sustentaculum" mentioned by Turner and Tchirkin (quoted by Hitchcock, 1940).

In advanced cases of spondylolisthesis, the body of the fifth lumbar vertebra actually hangs below the level of the anterosuperior aspect of the first sacral segment. In such cases an anteroposterior roentgenogram will reveal a so-called "bow-line" (Brailsford, 1929). This line may be simulated when the fifth lumbar segment is not displaced as far anteriorly, but in which a large bony excrescence forms between the anterior aspect of the fifth lumbar vertebra and the subjacent first sacral segment. In several of our cases this line involved only half of the anterior aspect of the upper sacrum as studied in the anteroposterior view, an observation which led to the identification of the large calcific spur as differentiated from the true bow-line of Brailsford.

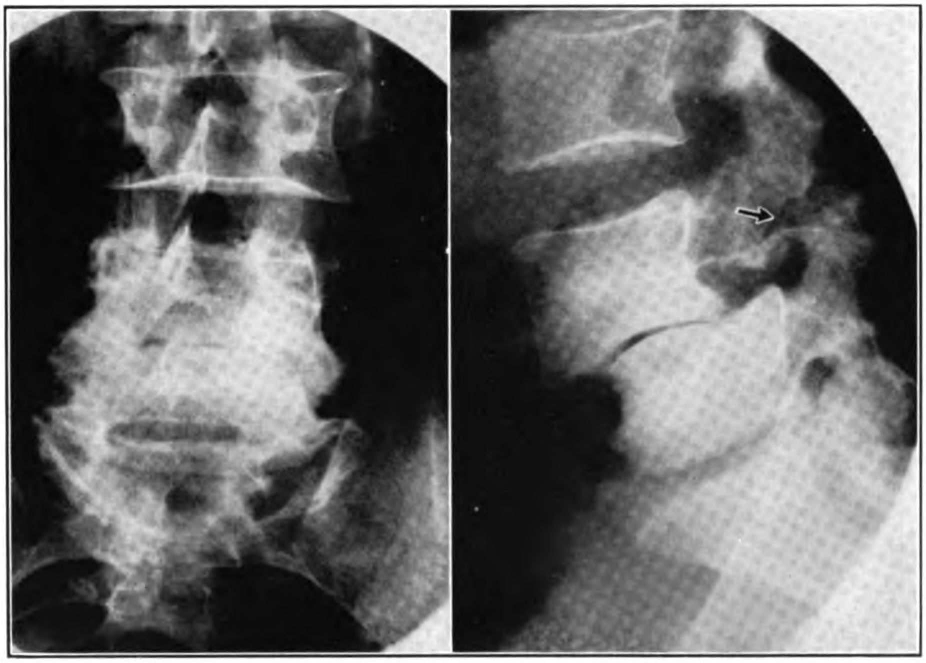

FIG. 271.—Grade II spondylolisthesis of the fourth on the fifth lumbar vertebræ, with ball and socket arrangement of the articular surfaces. The laminar defect is seen in the lateral view (arrows). Note the deformity of the intervertebral foramen. A mild reverse spondylolisthesis is present at the lumbosacral interspace.

Displacement of the fourth on the fifth lumbar vertebra rarely is as extensive as may occur at the lumbosacral articulation. I have not observed any instances in which the degree of such spondylolisthesis was more than grade 2, and most corresponded to grade 1.

One case of spondylolisthesis of the second on the third lumbar vertebra is of particular interest (Fig. 272). An anomaly of the lamina of the second lumbar vertebra was present, as evidenced by the diminished size of its

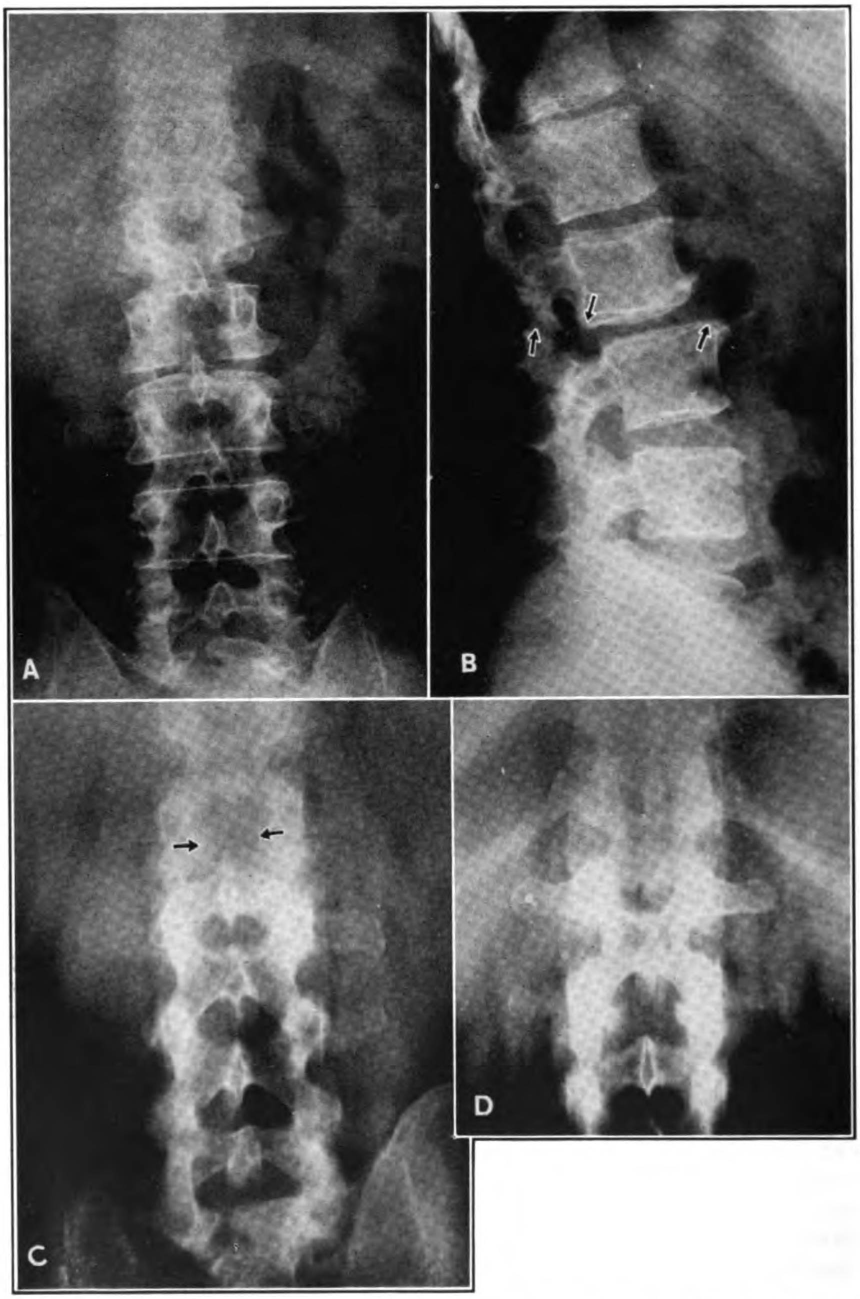

FIG. 272.—*A* and *B*, Reverse spondylolisthesis of the second on the third lumbar vertebræ (arrows). A congenital defect of the neural arch of the second lumbar vertebra is present. *C* and *D*, Same case, body section roentgenograms of the neural arches showing defect (arrows).

spinous process and neural arch. This was best brought out on body section roentgenograms. In addition, a lack of fusion of the pars interarticularis of this vertebra was observed on the lateral views, the latter change being unidentifiable in the oblique roentgenograms.

Guri (1946) in a review of 151 cases of painful spondylolisthesis, reported that 137 of these involved the lumbosacral articulation. Of these 102 belong to grade 1, 16 to grade 2, 10 to grade 3 and 4 to grade 4. In addition, in 3 cases the displaced vertebræ were inside the pelvis in front of the first or second sacral segment. In the grade 1 spondylolisthesis 56 cases showed that the intervertebral disc was apparently normal. In 12 there were thinning of the posterior aspect of the disc while in 34 the disc was markedly or totally destroyed. Sclerosis of the bony surfaces was observed in most of the cases with disc degeneration. In others, a buttress formation was present at the anterior aspect of the upper sacrum.

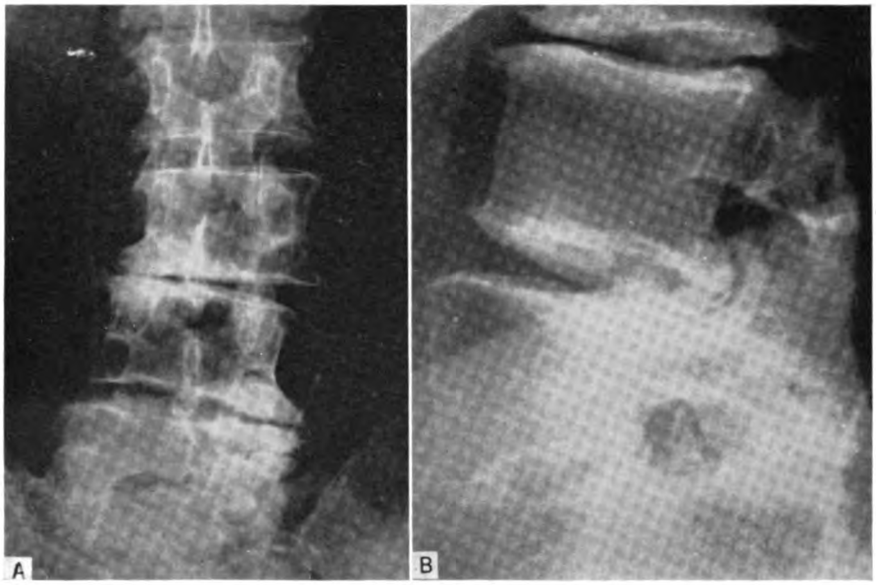

FIG. 273.—Lateral displacement and reverse spondylolisthesis of the fourth on the fifth lumbar vertebræ. The intervertebral disc is narrowed on its left side, and a spondylotic reaction is present. Note the narrowing of the third lumbar interspace, with the right side of this interspace involved to a greater extent than its left side. The lateral view (*B*) shows a posterior slipping of the fourth lumbar vertebra. The articular facet of the subjacent vertebra has become cupped concave anteriorly, and the intervertebral foramen is constricted by the displaced vertebra.

Practically all of the cases of grade 2 spondylolisthesis had localized spondylitis and thinning of the intervertebral disc of varying degree. In the cases of grade 3 and 4 spondylolistheses, which were subdivided into two groups according to whether or not new bone formation was present, a buttress at the anterior aspect of the sacrum was present in most cases. In others the sacrum and the body of the fifth lumbar vertebra were fused together. When new bone formation did not take place, rounding off of the anterior border of the sacrum and occasionally arching of the inferior surface of the fifth lumbar vertebra occurred, forming a joint-like image between the last lumbar vertebra and the sacrum.

With spondylolisthesis of the fourth on the fifth lumbar vertebra, localized arthritis was less than in lumbosacral spondylolisthesis. However, thinning of the intervertebral disc was not uncommon.

Arthritic changes occur between the articular facets of the lumbosacral articulation, and less frequently, in those between the fourth and fifth lumbar vertebræ. In addition, in the oblique roentgenograms one may sometimes identify increased bony densities at the margins of the defect in

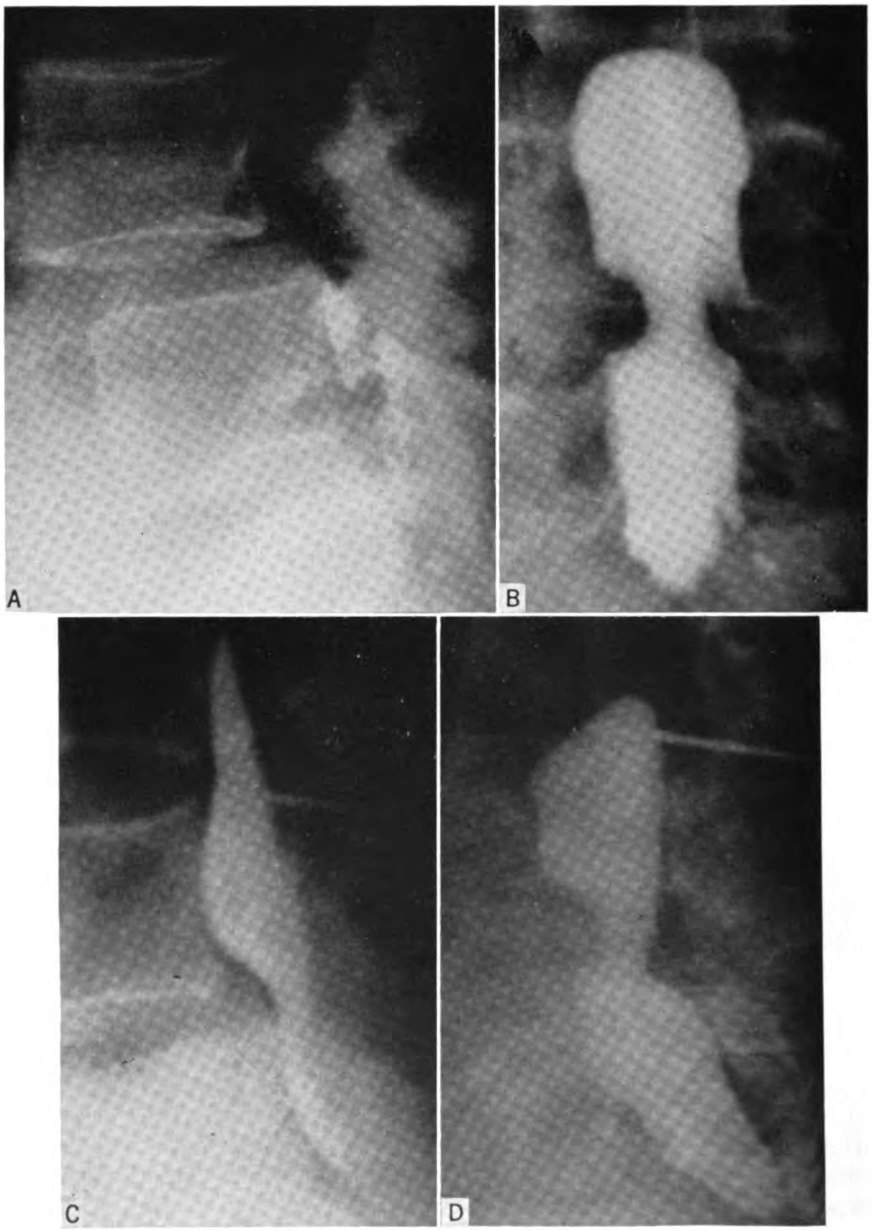

FIG. 274.—Symptomless grade I spondylolisthesis of L 4 on L 5. Myelogram performed because of symptoms relating to cervical pain. Note the waist-like defect (*B*) and the step deformity (*C*) and (*D*).

the pars interarticularis and in occasional instances small spur-like formations at these margins may be observed. It is possible that arthritic changes at the apophyseal joints of the fourth and fifth lumbar vertebræ afford an additional cause for low back pain. I believe that this occurs in a fairly large percentage of cases.

The appearance of the intervertebral disc with spondylolisthesis is worthy of comment. In early cases of grade 1 spondylolisthesis, the disc frequently is perfectly normal. However, with wear and tear, progressive degeneration of the annulus fibrosus and disruption of the fibrocartilage is followed by narrowing. This is first manifest in the posterior aspect of the intervertebral disc. Later, the disc may become reduced to only a thin wafer-like structure. It is believed that discal degeneration is one of the major factors in the progressive slipping of the involved vertebra.

In discussing cases of spondylolisthesis which have come to surgica treatment, one is impressed with the observation that there is great variability in the degree of fixation of the supposedly separated neural arch. In some cases my orthopedic colleagues have mentioned that the neural arch was readily moved about after the spinous process had been exposed surgically, and this was believed to be an indication of instability of the underlying vertebra as well. In other instances it was the opinion of the operating surgeon that relatively little, if any, instability existed because traction and pulling on the exposed spinous process barely moved the neural arch at all. Woolsey (1954) reported relief of pain in 7 cases of spondylolisthesis after surgical excision of the movable neural arch. Todd and Gardner (1958) favor simple excision of the unattached lamina as the operation of choice, and regard fusion as unnecessary.

It is well known that many individuals have a first degree spondylolisthesis with no pain whatsoever, and that following trauma or prolonged strain symptoms may become apparent and aggravated as the degree of instability of the lumbosacral spine increases. A radiologic picture resembling that of spondylolisthesis occurs in individuals with fractures of the pedicles and laminæ. One of our patients fell off a scaffold and suffered fractures of the laminæ of the fourth lumbar vertebra. Soon thereafter he developed a grade 2 spondylolisthesis. It was postulated that this patient probably had a weakened intervertebral disc which facilitated the gliding of the fourth over the fifth lumbar vertebra. The fifth lumbar interspace was narrowed, and there was posterior displacement of the fifth on the first sacral vertebra measuring 0.5 cm. The latter probably predated the injury. Another seven-year-old boy who fell under a roller coaster suffered a fracture of the pars interarticulasis of the fourth lumbar vertebra, and had a grade 2 spondylolisthesis.

Spondylolisthesis is accompanied by deformity of the spinal canal varying mainly with the extent of slippage. With grade 1 spondylolisthesis there is only a slight angulation of the anterior aspect of the dural sac as it passes over the lip of the subjacent vertebra. Little deformity occurs in the lateral aspects of the pantopaque column, and the axillary pouches are not disturbed. With further forward displacement a more pronounced step deformity appears in the anterior aspect of the pantopaque column, and its dorsal aspect becomes deformed in an opposite direction because of pressure from the neural arches. This results in a waist-like deformity, with a relatively narrow channel at the level of the involved intervertebral disc. In the frontal projection a variable bilateral indentation into the pantopaque column is apparent. It is important to realize that spondylolisthesis producing such a deformity of the available space may exist without symptoms. On the other hand, even small intrusions into the spinal canal at the level of the spondylolisthesis can cause marked compressive changes on the cauda equina and nerve roots, with consequent clinical manifestations. The association of herniations of the intervertebral disc with spondylolisthesis is infrequent. The compression of the cauda equina and nerve roots at the

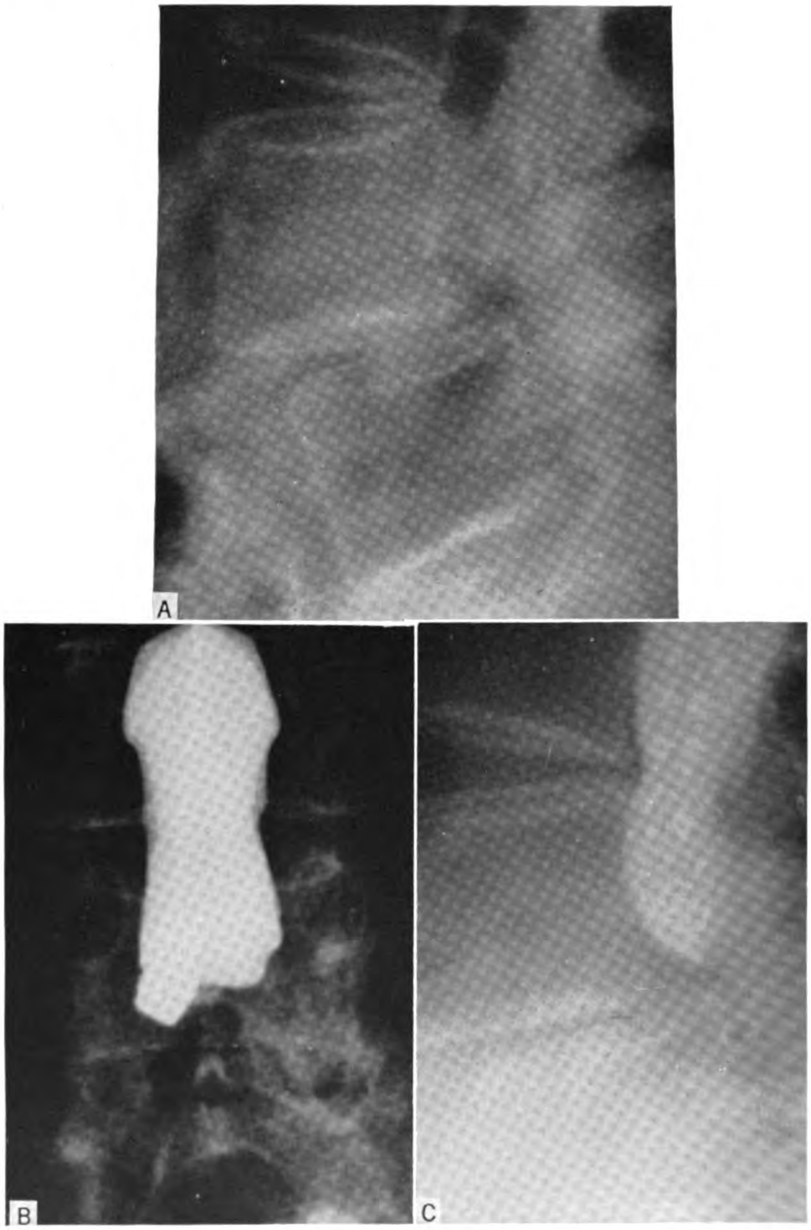

FIG. 275.—Grade II spondylolisthesis in patient with bilateral sciatic pain and pain in the lower back. The intervertebral disc is reduced in height and the intervertebral foramina are encroached upon. A complete myelographic block was found (*B* and *C*). In addition to the spondylolisthesis a definite ridge was encountered together with only a modest discal herniation. The combination of spondylolisthesis, ridge and disc deformities caused the block.

level of the spondylolisthesis is due mainly to pinching of the dural envelope by the displaced vertebra. This is heightened by the presence of a narrow spinal canal, spondylotic ridges, thickening of the ligamentum flavum and any hypertrophic change present at the region of the cleft in the pars interarticularis or thickening of the neural arch. Preoperative myelography is advisable if there is question as to caudal sac compression.

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Reverse Spondylolisthesis

In this condition a posterior displacement of the body of a lumbar vertebra on the vertebra beneath results from an anatomic variation in the articular facets between these structures. As a rule, the displacement occurs at the lumbosacral joint, and is only moderate. It is associated with downward and posterior alteration in the alignment of the lumbosacral articular facets. As originally reported by Johnson (1934), there were 12 such cases out of 126 with spondylolisthesis observed by him. He estimated the incidence of this condition as about 10 per cent. Seven of his 12 cases had narrowed intervertebral discs. Smith (1934), who reported the same condition, also believed that the displacement was the consequence of anatomic variations in the lumbosacral articular facets. Both agreed that if the degree of displacement was sufficient, sciatica might result from nerve root pressure. Smith, who reported 56 cases, mentioned that trauma played a secondary role. Degeneration of the intervertebral disc as well as loosening of the associated ligaments and frequent small traumas were considered by De Veer (1935) as important in the development of reverse spondylolisthesis. He, too, concurred that apophyseal joint derangements were the important factors in the origin of this condition.

An interesting group of 77 cases of vertebral retroposition was reported by Gillespie (1951). These were encountered in a series of 493 cases of lumbosacral disc protrusions confirmed at operation. All involved the lumbosacral articulation. Thirty-five cases had more detailed examinations, and it was found that the average backwards displacement ranged from 0.3 to 0.9 cm. The majority of his cases showed an anteroposterior type of articular facet. Gillespie noted that retroposition may occur at any level, most often the lumbosacral and the cervical regions. The condition may occur with arthritis, protruded discs, degeneration of the intervertebral cartilages, infection and trauma. He did not consider it a cause of backache.

Fletcher (1947) noted a high incidence of degenerative changes in the posterior fibers of the annulus fibrosus as well as advanced degenerative disc disease with this condition. He stressed the importance of weakening of the intervertebral disc in the genesis of retroplacement of the fifth lumbar vertebra. He noted that there was no exaggeration of the lumbosacral angle, and that the apophyseal joints were principally of a frontal type directed downwards and backwards. In his opinion disease of the fibers of the annulus fibrosus came first, followed by posterior displacement as a mechanical consequence.

It has been mentioned that improperly positioned films of the lumbosacral articulation may simulate reverse spondylolisthesis. Willis (1935) reported on the examination of 50 consecutive museum skeletons. From analysis of the apposing lumbar and sacral surfaces he noted that a frequent difference existed in the anteroposterior depth of the fifth lumbar vertebra and the first sacral vertebra. The anteroposterior diameters were equal in 34 per cent of his cases, while the remaining 66 per cent showed that the sacral diameter was from $\frac{1}{16}$ to $\frac{1}{4}$ inch shorter than the lumbar diameter. Posterior displacement of the lumbar segment could therefore be attributed to the relative shortening of the sacral diameter. This posterior overlap was illusory, and was not a cause of pain. Melamed and Ansfield (1947) also found that technical errors might simulate this condition. Nevertheless, they considered it a true pathologic entity, the origin of which may lie in degenerative processes, trauma, various inflammatory diseases or congenital anomalies. In this connection the comment of Roche and Bryan (1946) that a reverse spondylolisthesis in the presence of an intact neural arch and posterior articulation may be the result of congenital shortening of the pars interarticularis is of interest.

Posterior displacement of a vertebra was mentioned as an early sign of tuberculous spondylitis of the lumbar spine by Hagelstam (1947). He observed that laxity of vertebral fixation which results from destruction of a disc permits slight displacement of the vertebra in the horizontal plane. A retroposition of less than 1 cm. of one or more vertebræ appeared in 5 out of 45 patients with lumbar spine tuberculosis. In all cases except 1 a narrowing of the intervertebral disc also was present. In 1 case of tuberculosis in an osteoporotic spine the vertebral retroposition was the only diagnostic radiographic sign. Hagelstam also reported a case in which the causative disc lesion was due to staphylococcic infection.

We also have observed reverse spondylolisthesis occasionally. In most cases this was unassociated with pain which might be attributed to the condition, and as described by others, was accompanied by narrowing of the intervertebral disc and coronal position of the articular facets. One case was of particular interest. This was a sixty-seven-year-old woman in whom reverse spondylolisthesis was present at the level of the fourth and fifth lumbar vertebræ, with a posterior displacement of the fourth of 1.4 cm. The condition was identified on a lateral film of the abdomen made during the course of a gastrointestinal examination. On the anteroposterior view a marked narrowing of the third and fourth lumbar intervertebral discs was seen. A heavy bony spur was present on the inferior surface of the third lumbar vertebra and the superior surface of the fourth on the right side. An equal degree of narrowing was noted at the fourth lumbar interspace on the left side. On lateral views the lumbar lordotic curve was straightened. In addition to the reverse spondylolisthesis at the fourth interspace, a slight posterior displacement of the third on the fourth lumbar vertebra was present. The articular facets between the fourth and fifth lumbar vertebræ as seen on the lateral view were concave anteriorly. Those between the third and fourth lumbar vertebræ were atypical in that the right apophyseal joint was partially anteroposterior in configuration. The rotation of both the third and fourth lumbar vertebræ occurred on the coronally placed apophyseal joints.

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Pseudospondylolisthesis

This condition is predicated upon displacement of a vertebral body anteriorly because of weakening of the apophyseal joints producing a forward inclination of the inferior articular facets, with consequent forward displacement of the suprajacent vertebra. The neural arch remains intact, and no isthmic defect is present. The condition is usually accompanied by spondylosis and weakening of the intervertebral disc. It occurs more often in females, and the usual site is at the fourth or fifth lumbar vertebræ. Although brought into prominence by Junghanns (1931), its existence had been known for a long time (Friberg, 1939). Guri (1946)mentioned that according to Sisefski the term "pseudospondylolisthesis" was first used by Meugebauer for cases of violent vertebral dislocations secondary to fracture.

In a collection of Eskimo and Indian skeletons (Stewart, 1935) found 3 specimens with first or second degree spondylolisthesis without separated neural arches. This was shown to be due to arthritic erosion of the articular facets. Macnab (1950) reported 22 such cases in a series of 142 patients with spondylolisthesis. The roentgenograms in this series revealed an increase in the angle between the articular facets and the pedicles which permitted subluxations at the inferior joints. The anterior displacement averaged less than 1 cm.

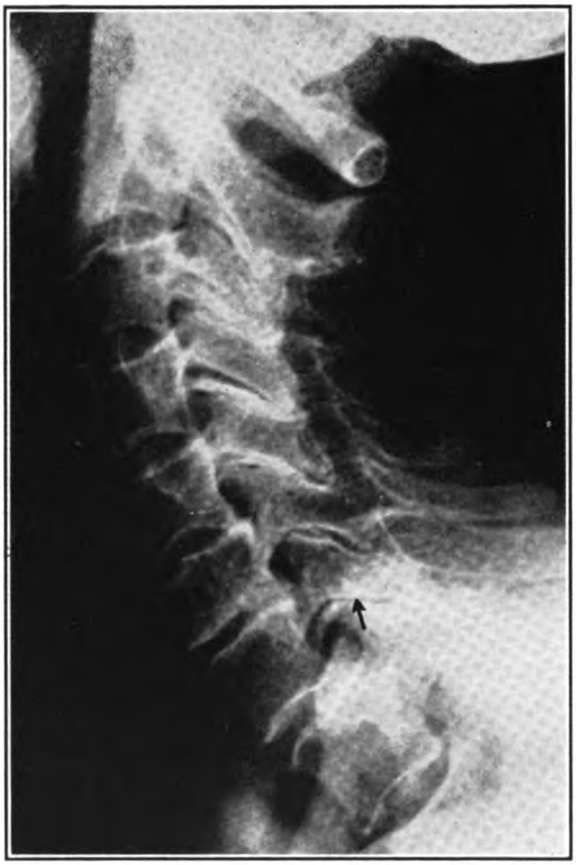

FIG. 276.—Pseudospondylolisthesis of the sixth on the seventh cervical vertebræ, with forward inclination of the articular facets (arrow).

Frequently identified with pseudospondylolisthesis was a clinical syndrome of backache and sciatica. Macnab also observed an increased range of motion of the vertebral bodies with flexion and extension. Degenerative discal changes, spondylosis and osteoporosis were also described, as well as the arthritic changes in the apophyseal joints.

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CERVICAL SPONDYLOLISTHESIS

A case of spondylolisthesis of the sixth on the seventh cervical vertebra was reported by Perlman and Hawes (1951) in an adult patient with bilateral pedicle defects. Kornblum, Clayton and Nash (1952) observed that the acute stage of rheumatoid arthritis might be accompanied by hyperemia and capsular distention of the cervical joints, together with deforming muscle spasm. This may produce dislocations of the various cervical segments. They reported 4 cases to stress that lower cervical spine as well as atlanto-axial dislocations may occur with this condition. In 1 case (case 3) they described a slight anterior dislocation of the fifth on the sixth cervical segment. The initial displacement was only 2 or 3 mm., but six months later it was a full 2 cm. forward, with about a 30-degree kyphotic and right lateral angulation and marked encroachment on the neural canal. Wide separation of the apposing articular surfaces of the posterior articulations at this level were noted, with marked erosion of the superior articular facets of the sixth vertebra. This was reduced by skull traction, and repeat roentgenograms one year later showed only minimal posterior displacement of the fifth cervical vertebra, with narrowing of the fifth cervical interspace. The articular facets were recalcified, with partial fusion of their surfaces.

We have noted that advanced cervical spine spondylosis, with increased cervical lordosis and prominent dorsum rotundum not infrequently is accompanied by a slight but fairly definite forward slip of the fifth or sixth cervical vertebra. This represents the cervical equivalent of the pseudospondylolisthesis seen in the lower lumbar spine.

In our cases the condition was asymptomatic. However, in patients with a considerable spurring along the dorsal aspect of the discs such a slip might prove important when symptoms of cord pressure appear. The so-called ridge syndrome of spondylosis may be heightened if a forward displacement of one of the lower cervical segments appears.

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UNCOMMON FORMS OF VERTEBRAL DISPLACEMENT

Displacement of the vertebral body in cretinism occurs occasionally when there is hypoplasia and posterior displacement of the first lumbar body. However, this does not qualify as a true displacement, but rather is an unstable arrangement of the lumbar vertebræ with an incomplete development of the anterior aspect of the involved vertebra, giving rise to a gibbus-like configuration. Similar changes may be observed with gargoylism and achondroplasia.

Maldevelopment of the vertebral bodies and their neural arches with multiple defects, such as multiple congenital hemivertebræ, also may produce various forms of vertebral displacement.

Displacement of vertebræ because of metastatic disease may sometimes be observed. An example of this is included here, in which myelomatous destruction of the neural arch and vertebral body resulted in a dorsal displacement of the first lumbar vertebra (Fig. 273).

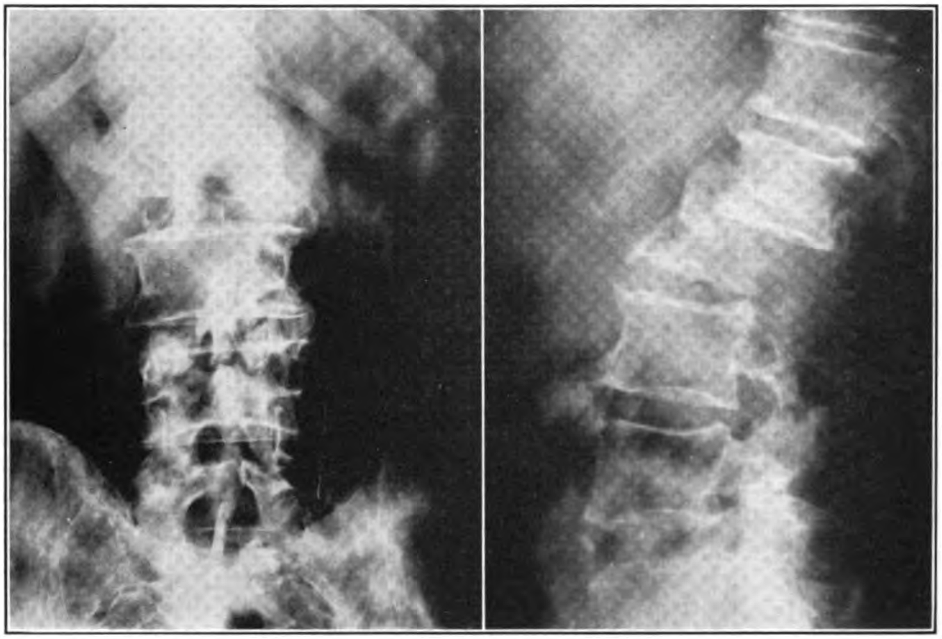

FIG. 277.—Retrodisplacement of the first lumbar vertebra due to myelomatous destruction of the second lumbar vertebra.

PSEUDARTHROSIS

In view of the frequency with which spinal fusion is employed for the relief of spondylolisthesis, a method for checking the effect of the operation is of interest. Cleveland, Bosworth and Thompson (1948) found that pseudarthrosis could best be demonstrated on films made in flexion and extension, and with bending to the right and left. Superimposition of identical films was the best way of evaluating untoward movement. Thompson and Ralston (1949) used flexion and extension films, but mentioned that at times the condition could be diagnosed only at operation. In the present author's experience body section roentgenograms through the fused areas have sometimes been helpful. At best, however, these methods are of doubtful reliability.

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Chapter

7

Diseases of the Intervertebral Discs

JUVENILE KYPHOSIS DORSALIS (SCHEUERMANN'S DISEASE)

This condition was first described by Scheuermann (1936) as an osteochondrosis, predominantly of the middle, lower thoracic and the upper lumbar vertebræ. He ascribed this to deficient epiphyseal growth of the vertebræ, occurring more often in females between the ages of ten and fourteen years. Schmorl objected to the term "vertebral epiphysitis" as designating the apophyseal line as an epiphyseal line, inasmuch as it is not a true growth epiphysis. Schmorl pointed out that vertebral growth occurred on the spongiosa side of the cartilage end plates, which are the true growth zones. He held that weakening and thinning of this growth zone together with herniations of the intervertebral disc, with resultant decrease in the anterior height of the vertebral body, was the primary cause of this disease. It has been established that the ring apophyses play no part in the growth of the vertebral body, and that disturbances in this structure visualized roentgenologically are the result rather than the cause of juvenile kyphosis.

MacGowan (1944) pointed out that with adolescent kyphosis the nucleus pulposus is disrupted before ossification of the vertebral body is complete. This destruction may be caused by failure of the cartilaginous disc to retain the nucleus, or by softening of the vertebral body. When this happens the weight-bearing function is transferred to the vertebral bodies anteriorly and the axis of motion to the posterior vertebral elements. Increased pressure results at the anterior borders of the vertebral bodies when the spine is in flexion. If the patient is adolescent, the epiphysis will be subjected to trauma and abnormal ossification follows. Premature closure of the epiphyses, failure of anterior vertebral ossification and wedging of the bodies constitute the end results. MacGowan's experience differed from others in that his patients were predominantly male. Knuttson (1948) studied juvenile kyphosis by means of serial films of growing children. He concluded that this disease retarded vertical growth of the vertebral bodies involved. This resulted in an intact rectangular shape with diminished height in some, while in others narrowing anteriorly produced wedge-shaped deformities. He considered Scheuermann's disease as an obscure disturbance of growth involving either the entire surface or only a portion of the vertebral body.

The association of spinal extradural cysts and kyphosis dorsalis juvenalis was described by Cloward and Bucy (1937). These authors believed that the changes in the vertebræ were not of an inflammatory nature, and did not predominantly or selectively involve the vertebral epiphyses. They stated that the many vascular channels in the epidural space comprising the vertebral circulation may become occluded by the pressure of a cyst wedged between the bony walls of the spinal canal and the dura mater. The changes in the vertebral bodies could then be explained on the basis of venous congestion. The partial collapse of the contiguous vertebral bodies resulted in the rounded and fixed deformity. They found this to be self-limited and never severe, assuming an arrested state within a few months after the onset. While this may occur in cases of spinal extradural cysts, which are not common, the mere frequency of juvenile kyphosis suggests that the mechanism described by Cloward and Bucy, is not a major factor in the production of the deformity. The concept that an osteochondrosis or aseptic necrosis constitutes the underlying cause likewise is not accepted.

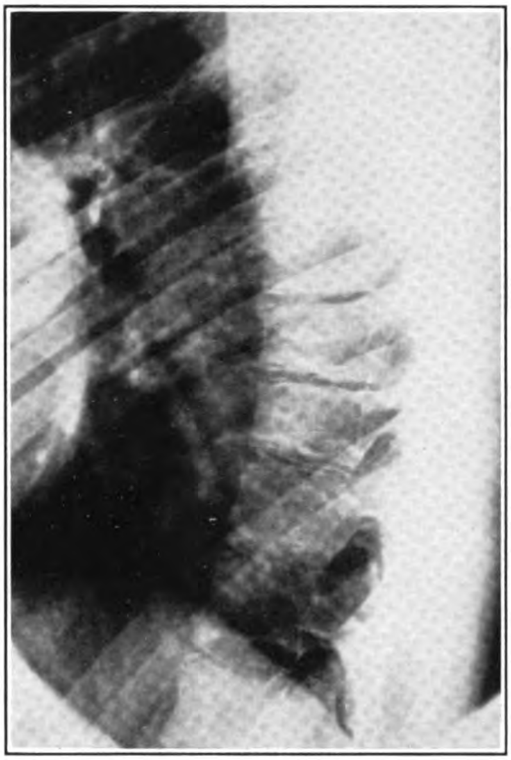

FIG. 278.—Early changes of Scheuermann's disease. Very little dorsum rotundum is present. The intervertebral spaces are narrowed, and the articular surfaces of the vertebræ are irregular. The ring apophyses are not fragmented at this stage. The midthoracic vertebræ are slightly wedged.

The clinical picture associated with juvenile kyphosis varies considerably. In the early stages of the disease the diagnosis may be made accidentally on the discovery of characteristic changes in the articular surfaces of the vertebral bodies on examinations made for other purposes. Symptoms may be mild or even absent during the acute stage of the disease. In others the disease manifests itself because of persistent mild pain between the shoulder blades associated with a stooping posture. There may be tenderness on pressure over the involved area and the pain may be aggravated when the patient has been erect or after exercise. Symptoms referable to compression of the spinal cord are not common, but Wretblad

(1939) reported 6 patients, all young men engaged in hard physical labor, who also had numbness, weakness, and unsteadiness in the legs. Spastic paralysis of the legs appeared in several cases. Myelograms in these individuals showed a block at the apex of the curve and improvement followed laminectomy. In 1 patient in whom symptoms had been present for several years and who died after laminectomy, autopsy showed irreparable damage in the spinal cord. Van Landingham (1954) reported a seventeen-year-old male in whom herniated discs in the mid-dorsal spine was associated with Scheuermann's disease. These were demonstrated myelographically, and the spinal cord compression had to be relieved surgically. More often myelography discloses only a gradual narrowing of the subarachnoid space at the apex of the curve (Lindgren, 1941).

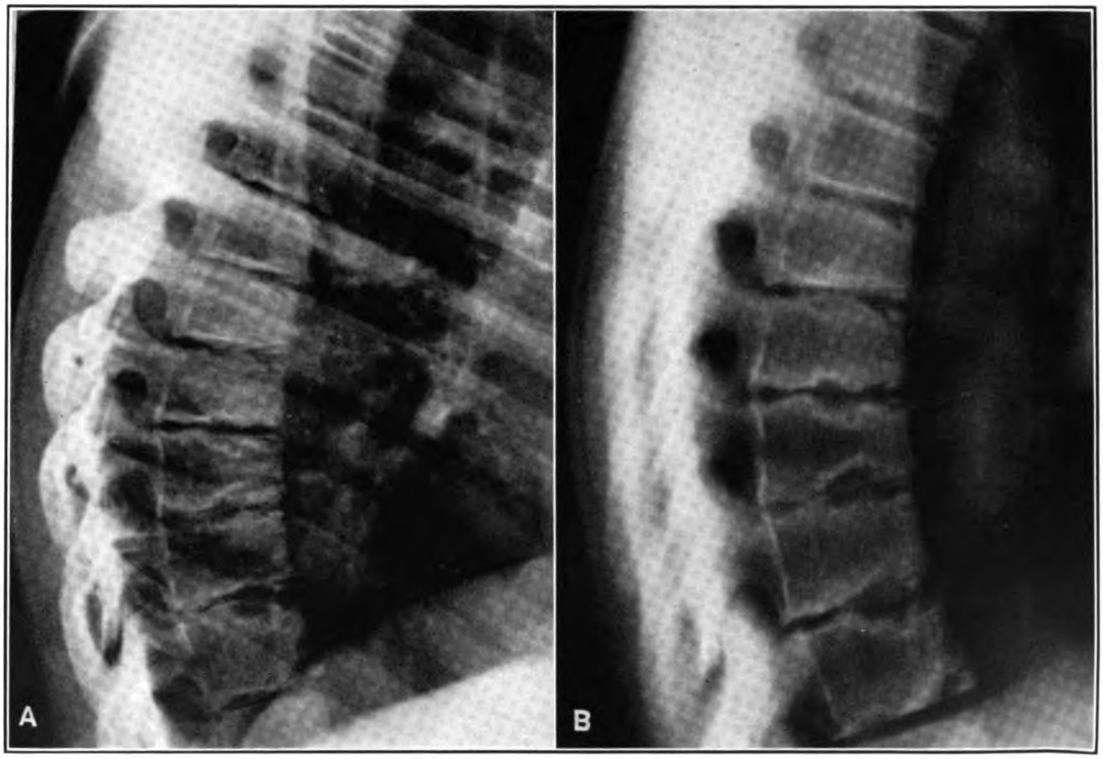

FIG. 279.—*A*, Moderately advanced case of kyphosis dorsalis juvenalis. Note the nuclear protrusions into the superior and inferior surfaces of the vertebræ. *B*, Same case, lateral body section roentgenogram. The nuclear protrusions with some bony reaction around them are well demonstrated. The ring apophyses in the midthoracic vertebræ are rather large but intact. In the lower thoracic vertebræ the ring apophyses are widened, irregular and have bony rarefaction immediately beneath them.

The radiologic appearance of juvenile kyphosis is variable, depending upon the stage of the disease in which the diagnosis happens to be made and the number of vertebræ affected. In the early stages of the disease the roentgenograms show only a slight irregularity of the vertebral end plates associated wih slight decalcification of the peripheries of the vertebral bodies immediately adjacent to the apophyseal plates. At this stage of the disease there may be no alteration in the usual alignment of the thoracolumbar spine.

As the disease progresses the irregularities of the plates become more marked and decalcification of the margins of the vertebral body appear.

The intervertebral discs between the involved vertebræ become narrowed and irregular with nuclear extrusions projecting through the end plates into the vertebral bodies. There may be a pronounced degree of decalcification in the anterior aspects of the vertebral bodies. In more advanced cases deficiency of bone appears with resultant irregular tongue-like projections of the anterior aspect of the involved vertebral body

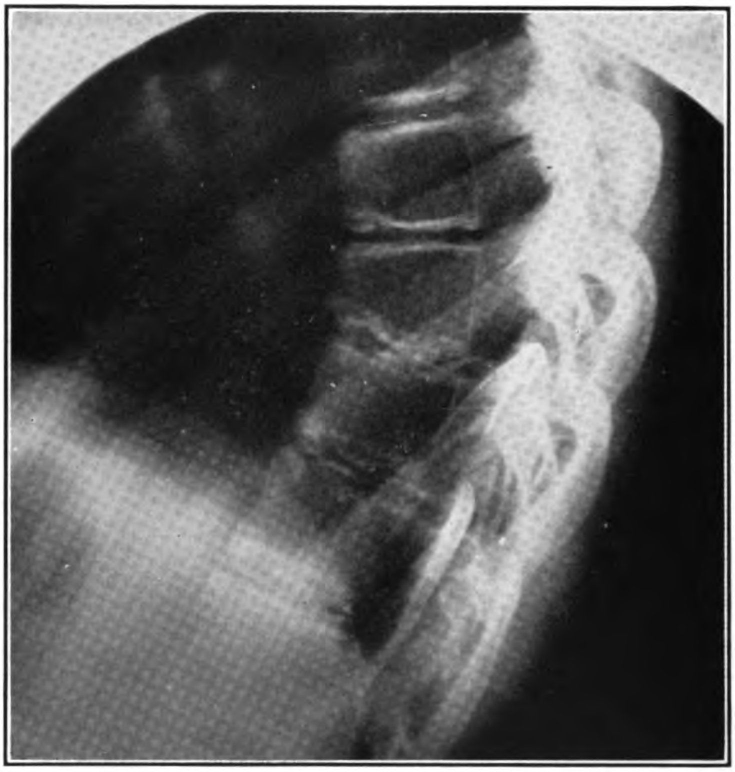

FIG. 280.—Kyphosis dorsalis juvenalis. Note the early wedging of the vertebræ at the apex of the curve.

Associated with the more advanced changes in the vertebral bodies are a progressive wedging and notching of the anterior aspects of the vertebræ, producing a kyphotic change. In the frontal plane scoliosis may be observed if the wedging of the vertebral body is irregular, leading to a diminished height on one side as compared with the other. As the disease passes into a healing stage, and eventual cure, the fragmented appearance of the end plates and the associated irregular decalcifications in the vertebral bodies disappear. By the age of eighteen to twenty years complete union of these elements occurs, resulting in a well-calcified vertebral body with an anterior wedge-like deformity. Persistent bits of calcification remaining at the anterior aspects of the vertebral bodies have been referred to as Scheuermann's nodes, and sometimes are mistaken for fractures. Similar bony changes occur in the lumbar and the cervical spine. Occasionally such ununited fragments of bone may be residual from the pathologic

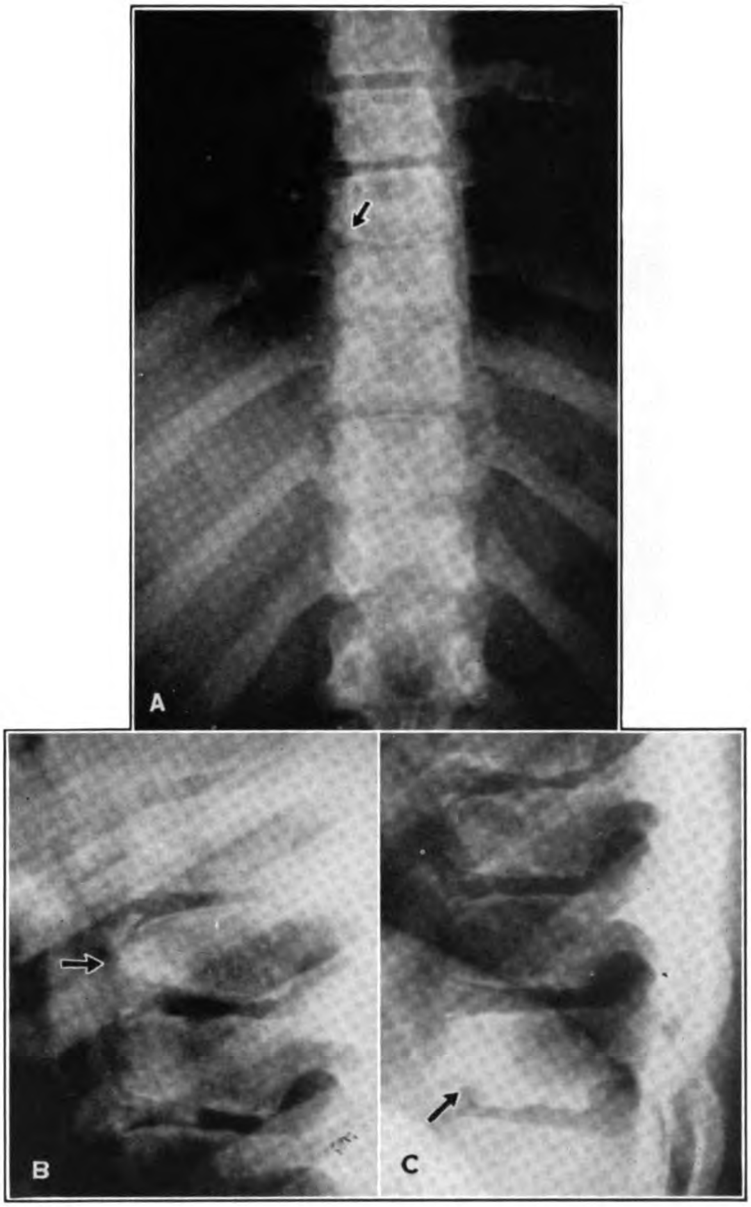

FIG. 281.—*A*, Anteroposterior view of thoracic spine in twelve-year-old girl with kyphosis dorsalis juvenalis. Note the blunting of the right inferior aspect of the eighth dorsal vertebra (arrow). *B* and *C*, Same case. There is wedging of the anterior aspect of the seventh thoracic vertebra (arrow) and excavation of the anteroinferior aspect of the body of the twelfth thoracic vertebra (arrow) by intervertebral discal herniation.

process associated with juvenile kyphosis. It has been established by discography that these bony changes are due to anterior discal herniations and represent bits of bone separated from the main body of the vertebra by intervening islands of cartilage.

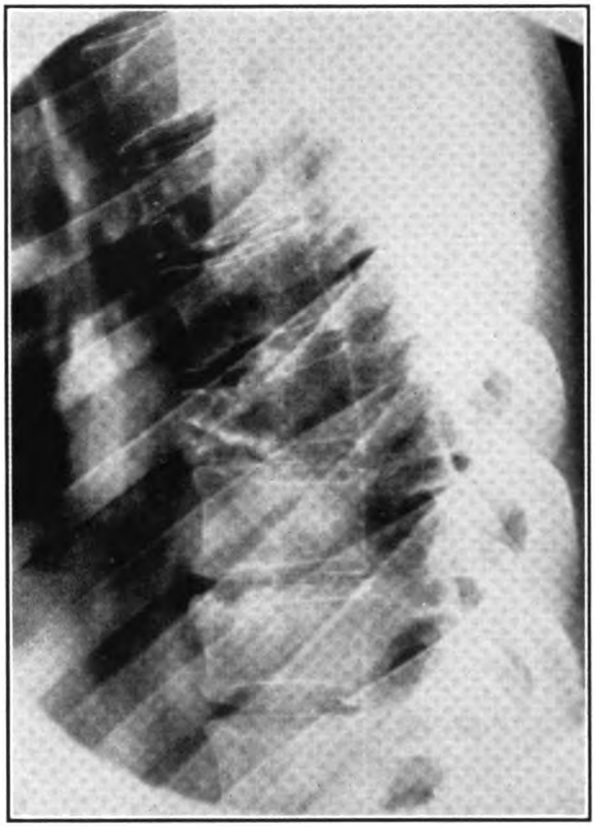

FIG. 282.—Kyphosis dorsalis juvenalis with pronounced wedging of the anterior aspect of the seventh dorsal vertebra. Note the nuclear impressions into the inferior aspect of this vertebra, and the relative absence of dorsum rotundum.

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INTERVERTEBRAL DISC INJURIES

Disc Puncture.—In view of the frequency with which lumbar puncture is performed for myelography and discography, it is worth reviewing the possible hazards of this procedure. It is known that perforation of the posterior spinal ligament may be followed by changes in the intervertebral disc. Pease (1935) observed discal narrowing after needle puncture through the posterior spinal ligament. Progressive narrowing of a lower lumbar intervertebral space and Schmorl's nodes were noted by Gellman (1940) on successive roentgenograms of a fourteen-year-old girl who had had a spinal tap followed by severe pain and lumbar hyperextension. Epps (1942) also reported a case of degeneration and collapse of an intervertebral disc following lumbar puncture, which he attributed to aseptic degeneration. Three similar cases were described by Everett (1942). Downing (1944) reported 2 cases in which the intervertebral disc collapsed following operation under spinal anesthesia. He mentioned that if the needle were inserted so that it did not enter the subarachnoid space, and was further advanced so that it penetrated the annulus fibrosus and entered the disc in its lateral aspect, escape of nuclear material sometimes was followed by degeneration and collapse of the disc. In his opinion the increased flexion position used for lumbar puncture increased the possibility of such injuries. In his second case discal collapse occurred in a period of not more than sixteen days, too short a time for purely degenerative changes and probably better explained by escape of nuclear component. Baker (1947) observed narrowing of an intervertebral disc in a four-year-old boy after a spinal tap. Severe backache and rigidity of the spine appeared within a few days after lumbar puncture. Serial roentgenograms showed relatively rapid narrowing of the intervertebral disc.

In an experimental investigation of intervertebral disc lesions in dogs, Key and Ford (1948) found that after the discs had been punctured once by a 20-gauge needle, an atypical protrusion of the nucleus pulposus occurred in one instance. Congdon (1952) punctured the intervertebral discs in 13 rabbits, and observed that this was followed by regeneration of nucleus pulposus cells which presented some resemblance to neoplasm. In rabbits which survived from six to over ten months after such puncture, gelatinous nodules of nucleus pulposus cells were found in the adjacent soft tissues. Dripps and Vandam (1952) concurred that trauma to the annulus fibrosus may be followed by herniation of the nucleus pulposus. They stated that they had seen gelatinous material drop from the needle which had entered the nucleus pulposus of a fourteen-year-old girl who moved inadvertently as the needle was being advanced during a spinal tap.

On the other hand, Gardner, Weiss, Hughes, O'Connell and Weiford (1952) asserted that the small bore of the needle used to perforate the posterior spinal ligament for discography was not likely to damage either the ligament, the annulus fibrosus or the nucleus pulposus. Erlacher (1952), who suggested a lateral approach to the nucleus pulposus, bypassing the neural sac, also was of the opinion that the small gauge of the needle used did not increase the danger of nuclear protrusion. Cloward (1959) reported that no case of rupture of a normal intact disc followed discography in his series of over 450 lumbar discograms over an eight year period, and he regarded the fear of injury to a normal disc by needle puncture as unfounded. De Seze and Levernieux (1952) reported that a review of 59 patients who had had discography showed that thirteen had collapsed discs attributed to necrosis due to puncture and instillation of contrast material.

Among the other hazards which may be encountered during lumbar puncture, but which are quite uncommon, are hemorrhage into the cauda equina (Hammes, 1920), the formation of epidural hematomas and meningeal irritations caused by entrance of blood into the spinal subarachnoid space. Damage to vessels accompanying nerve roots may produce ischaemia of these structures.

Our own experience in this respect is limited to patients who had spinal taps for myelography. Included in the approximately 3000 myelograms were several cases in which the needle had been forcibly pushed into the intervertebral disc because of sudden uncontrolled movements by the patient. None developed untoward reactions to this trauma. We have no personal experience with discography, because up to the present we do not feel that the procedure adds enough to diagnosis to warrant even the slight risk of multiple punctures.

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Disc Infections

Inasmuch as the normal intervertebral disc is avascular, hematogenous infections are rare. However, extension of an inflammatory process from adjacent bone through a crevice in the cartilaginous plate may occur, and probably explains the rapid degeneration of disc tissue seen with extensive osteomyelitis of the vertebral column. In those instances in which some vascularization of the disc is present because of a degenerative process, it is conceivable that a hematogenous infection may occur. However, no such cases have been clearly identified. It is generally agreed that infection of the disc is incident to infection in the adjacent bone. From the radiologic point of view, it may be extremely difficult, if not impossible, to detect a minimal osteomyelitis, so that the initial radiologic change may be a progressively decreasing height of an intervertebral disc.

In tuberculosis of the spine the intervertebral discs are somewhat resistant, but eventually succumb to the destructive process. The progress of such destruction is variable, and may be accompanied by a wide variety of change in which bone destruction is first seen. Then, as the lesion progresses, various discal deformities appear as destruction advances. Finally the disc may practically disappear. At the end stages in favorable cases when bony fusion occurs, the intervertebral disc may no longer be identified. When a tuberculous focus is initially implanted adjacent to the anterior spinal ligament, the intervertebral disc may become involved relatively early by spread of the infection behind these ligaments.

Brucellosis may cause relatively rapid destruction of an intervertebral disc which may proceed to total disappearance (Di Rienzo, 1944).

Direct infection can be introduced into the intervertebral disc during lumbar puncture for diagnostic purposes or discography. Another more frequent association of intervertebral disc infection is that with surgery for herniated discs. The rate of progression of the infection varies considerably, and is preceded by severe lumbar pain for a few days to several weeks after operation. At first no specific changes can be identified radiologically. As the infection progresses the involved intervertebral disc becomes narrowed, and the bone on either side loses its trabeculation, assuming a washed-out appearance. It is presumed that the original infection in postoperative discal herniation patients is in the bone. Healing is slow and is accompanied by later development of peripheral osteophytes, mainly on the anterolateral aspect of the involved vertebræ.

A benign form of osteitis of the vertebræ in children was reported by Bremner and Neligan (1953). They described seven patients, all under three years of age, with fever, limp, stiff back with painful spasm and limited hip motion. Symptoms increased in severity for about five to six weeks, then became stationary or improved. Radiographic examination of the spine disclosed narrowing of the disc space, with erosion of the adjacent margins of one or both vertebræ. Recalcification was observed on serial study.

Ghormley, Bickel and Dickson (1940) described a presumable acute infectious lesion of the intervertebral discs, characterized by clinical signs of infection of a mild nature, with backache, spasm and tenderness over the affected areas. These authors stated that the intervertebral disc at first remained unaltered, but later diminished in height. The narrowing persisted for several months, and then bony proliferation appeared at the margins of the involved vertebræ. In some of their cases bony fusion occurred. In others paravertebral soft tissue thickening appeared. They believed that if osseous involvement became evident, osteomyelitis probably was the diagnosis.

In a discussion of intervertebral disc lesions in children and adolescents (Key, 1950), it was mentioned that low back pain and sciatica were rare. When low back pain did occur, it was attributed to an inflammation of the disc. The spine roentgenograms in these patients showed nothing abnormal, unless congenital abnormalities were present, in themselves not responsible for either low back pain or sciatica.

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reported that no case of rupture of a normal intact disc followed discography in his series of over 450 lumbar discograms over an eight year period, and he regarded the fear of injury to a normal disc by needle puncture as unfounded. De Seze and Levernieux (1952) reported that a review of 59 patients who had had discography showed that thirteen had collapsed discs attributed to necrosis due to puncture and instillation of contrast material.

Among the other hazards which may be encountered during lumbar puncture, but which are quite uncommon, are hemorrhage into the cauda equina (Hammes, 1920), the formation of epidural hematomas and meningeal irritations caused by entrance of blood into the spinal subarachnoid space. Damage to vessels accompanying nerve roots may produce ischaemia of these structures.

Our own experience in this respect is limited to patients who had spinal taps for myelography. Included in the approximately 3000 myelograms were several cases in which the needle had been forcibly pushed into the intervertebral disc because of sudden uncontrolled movements by the patient. None developed untoward reactions to this trauma. We have no personal experience with discography, because up to the present we do not feel that the procedure adds enough to diagnosis to warrant even the slight risk of multiple punctures.

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THE EFFECTS OF TRAUMA ON THE INTERVERTEBRAL DISC

Because of the elasticity of the intervertebral fibrocartilage and the cushioning action of the intact nucleus pulposus, the intervertebral disc usually is not affected by mild trauma. However, repeated or violent injury may produce rupture and fissure of the cartilaginous plates and consequent extrusion of nuclear material through such rents. Rotary or compressive trauma may injure the lateral aspects of the annulus fibrosus and be accompanied by fracture of the adjacent bone. Injuries to the posterior spinal ligament as a cause of posterior protrusion of the nucleus pulposus and discal material is a well recognized entity.

Progressive degeneration and narrowing of the intervertebral disc may follow extrusion of nuclear material. Part of the picture of healing is the development of osteophytes along the lateral and anterior margins of the involved vertebræ. Sclerosis of the apposing bony surfaces of a narrowed disc, particularly in the lower lumbar area, is frequent. Such changes were produced experimentally by Compere and Keyes (1933). They reported that in dog operations in which the intervertebral disc was opened and nuclear material curetted, the discs completely disappeared. The adjacent bony surfaces became markedly sclerosed, with spur formation and lipping of the anterior edges. These dogs were sacrificed three months after operation, and histologic study revealed that the discs at the site of injury had vanished. The bone had become dense, with marked hyperostosis and new bone formation similar to that seen clinically in cases of marked hypertrophic spondylitis.

With severe trauma to the spine, there may be a complete break through the fibrocartilage with shifting of the vertebral bodies for an appreciable distance. This is usually associated with fracture dislocations of the involved vertebræ. Wright and Gardner (1952) reported a case of traumatic chylothorax incident to dislocation of the sixth on the seventh thoracic vertebra. A complete transection of the spinal cord caused the death of this patient.

It is well known that in hyperflexion injuries of the neck retropulsion of the nucleus pulposus through the posterior spinal ligament into the spinal canal occurs, with consequent damage to the cervical cord (Gay and Abbot, 1953). The projected nucleus pulposus may return to its normal position. Such cases have been reported by Taylor and Blackwood (1948), Schneider (1951), and Cramer and McGowan (1944). It is worth recounting the case reported by Brooke (1944), in which a patient was admitted with a complete flaccid paralysis below the fifth cervical segment, with loss of sensation of all modalities below the sixth cervical segment. Necropsy fifteen days after admission showed defective and frayed posterior spinal ligaments, with posterior protrusion of a calcified nucleus pulposus which had impinged against the spinal cord, producing a soft defect in the lower cervical region. These authors noted that when the head was flexed slightly, this irregularly calcified intervertebral disc would bulge about 1 cm. into the spinal canal.

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DEGENERATIVE ALTERATIONS IN THE INTERVERTEBRAL DISC

Although degeneration of the intervertebral disc was considered in the chapter on spondylosis, the importance of these changes make it advisable to review briefly their anatomic and radiologic characteristics. The intervertebral discs are enveloped by the annulus fibrosus, which is thickest in its anterior portion where it blends with the anterior spinal ligament. The annulus is formed by fibrous and elastic lamellæ, with irregularly scattered cartilaginous cells attached to the adjacent bone by Sharpey's fibers. These are arranged in an oblique and spiral fashion, extending from one vertebral body to another, with considerable variation in the anatomic placement of their lateral aspects. The posterior portion of the annulus fibrosus is its weakest. In some instances the structure may be so poorly defined as to present breaks in continuity. Encased within the annulus fibrosus lies the nucleus pulposus, an elastic, gelatinous, almost lozenge-shaped structure which merges imperceptibly with the annulus fibrosus. In childhood and early adult life this structure is relatively smooth and homogeneous. Its turgor and elasticity are best during the early years of life. The cellular components of the nucelus pulposus are mostly fibroblasts and cartilage cells together with notochordal remnants. Above and beneath the nucleus pulposus, forming respectively the superior and inferior surfaces of the vertebræ are the cartilage plates. These are thickest centrally, and merge with the annulus fibrosus peripherally. Under normal circumstances the plates are complete, but small crack-like defects are quite frequent. Through these perforating fissures may project cancellous bits of bone, or fragments of the nucleus pulposus may enter into the vertebral bodies. Normally the disc is avascular, receiving its nutrition by means of perfusion, a process aided by the presence of lymphatics at the junction of bone and cartilage. With advancing age and in the presence of degeneration, some degree of vascular intrusion may appear.

Although degenerative changes of the intervertebral disc have long been known, today's knowledge stems mainly from the work of Schmorl and his co-workers. This was recounted in detail in the classical monograph by

Schmorl and Junghanns (1957, fourth edition). An excellent brief review of the normal and pathologic intervertebral disc was published by Coventry, Ghormley and Kernohan (1945).

Progressive degenerative structural changes of the intervertebral discs are closely related to fluid loss, with consequent alterations of turgor and elasticity. The hydration of intervertebral discs is greatest in the neonatal period, and diminishes slowly thereafter. A relatively more rapid loss of fluid content is noted for a short time during infancy. Thereafter the rate of dehydration is relatively slow until about the third decade, when the process of dehydration becomes accelerated. The borderline between the normal and abnormal at this stage is incident to the wear and tear of living. With advancing degeneration of the intervertebral disc one notes a gradual dissolution and atrophy of the annulus fibrosus with progressive softening and weakening of its structure. Necrotic changes may appear in the anterior portion of the annulus fibrosus weakening the ring and permitting prolapse of nuclear material. The nucleus pulposus likewise loses its gelatinous turgor because of loss of fluid, and in association with weakening of the annulus fibrosus and changes in the cartilaginous plates, the lozenge shape of the nucleus pulposus alters. These changes were well described by Erlacher (1952), who correlated the anatomic and radiologic appearance of the nucleus pulposus by means of a staining technic combined with radiography. He identified five principal forms, including a normal globular nucleus, a lobed nucleus with a few large lobulations which he considered normal in adults, a simple branched nucleus with long narrow limbs, which appeared to be predisposed towards herniation. He also mentioned a multiple branched nucleus with a small central shadow and several extensions considered a transitory form of degeneration in which prolapse was likely to occur. The fifth form was the spread nucleus with many branches in all directions instead of a single central shadow. This last type represented a typical degenerated disc, and was most often associated with a narrow intervertebral disc space. In such an instance, herniation was no longer anticipated.

An integral part of the process of degeneration takes place in the cartilaginous plates. In the earliest stage, fissures appear in the plates permitting protrusion of small bits of nucleus pulposus. With advancing changes, sometimes precipitated by injury, fractures of the end plates may appear and the nuclear material protrudes in various directions. Detailed studies of such protrusions with perforation of the cartilaginous plates appear in the work of Schmorl. As a rule, escape of nuclear material into the cancellous bone above and beneath the nucleus pulposus is not associated with clinical manifestations. However, if narrowing of the disc is sufficient to distort and diminish the intervertebral foramina, pressure may be exerted against the respective nerve roots.

As mentioned before osteophytes may be produced as a healing mechanism for degeneration of the intervertebral disc. Such bone formations are induced by thinning of the disc, and originate at the insertion of the longitudinal ligaments (Schmorl, 1932). Pain in the back and limitation of vertebral movements are rather infrequent in association with degenerative lesions of the intervertebral discs. However, with involvement of the apophyseal joints back pain may become apparent.

The association of changes in the intervertebral discs with alterations in spinal curvature is exemplified by the condition known as adolescent kyphosis. The etiology of this is still unknown. It is characterized by changes in the intervertebral discs with protrusions of the nucleus pulposus particularly into the anterior aspects of the vertebral bodies. According to Saunders and Inman (1939) it was described by Schanz in 1907. Nevertheless, it is through the work of Scheuermann (1921, 1936) that this condition was generally recognized. It was Scheuermann's opinion that the alteration was due to an epiphysitis analogous to Legg-Perthes disease. However, Schmorl (1930) and others preferred an explanation based on the necropsy observations of gross nuclear escape. From the roentgenologic point of view, the usual changes associated with Schmorl's nodes are said to be absent from adolescent kyphotic changes because of lack of sclerotic reaction about the prolapsed bits of nucleus pulposus. On laminagraphic examinations made in a few cases we have been able to demonstrate that sclerotic changes do occur (Fig. 279).

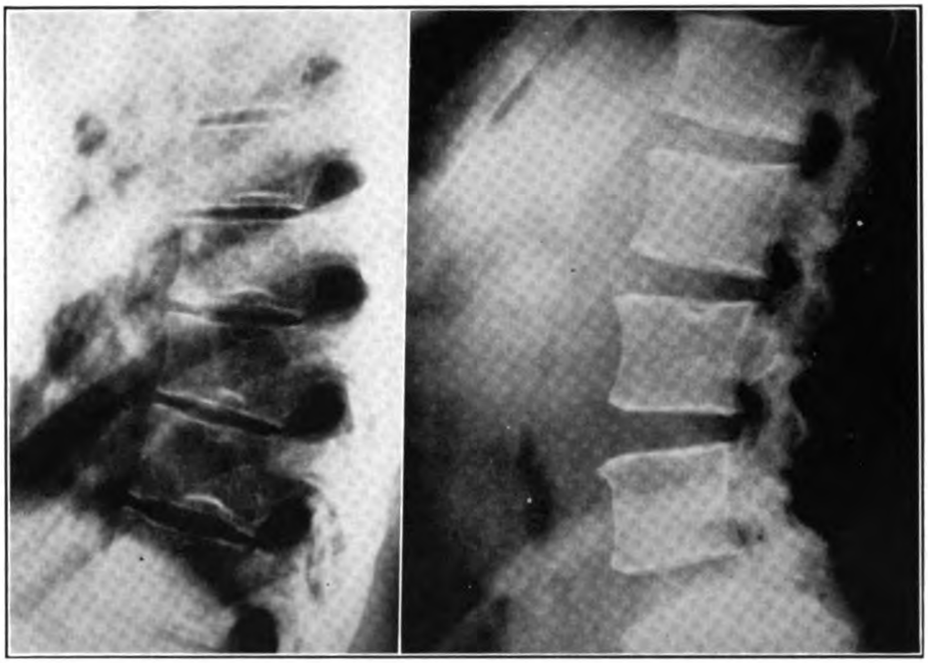

FIG. 283.—Thoracolumbar spine with multiple Schmorl's nodes in an athletic, nineteen-year-old girl who was symptom free.

Senile kyphosis occurs as a curvature of the upper and middle portions of the dorsal spine which appears late in life (Fig. 95). This condition is attributed to degenerative changes principally in the anterior aspects of the involved intervertebral discs, which assume a tapered appearance with the narrow end directed anteriorly. With progressive narrowing, the edges of the vertebræ may eventually touch, and actual bony fusion occurs. The posterior aspects of the discs may be narrowed, but usually remain identifiable even with advanced senile kyphosis. The vertebral bodies become cuneiform in shape, with some osteophyte production at the lateral aspects. It is not uncommon to observe a considerable degree of osteoporosis with senile kyphosis. Interestingly, the wide biconvex appearance of the discs associated with the swelling of the nucleus pulposus usually seen with osteoporosis does not occur when senile kyphosis is present.

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CALCIFICATION OF THE INTERVERTEBRAL DISCS.

According to Sandström (1951) calcification of the intervertebral discs was mentioned by Luschka (1858), and were first demonstrated roentgenologically in cadavers by Beneke (1897). In 1930 Calvé and Galland described them during life. The pathologic aspects of the condition were investigated by Schmorl (1929), who believed that calcification followed small discal ruptures followed by necrosis. Other etiologies proposed included persistent overloading of the spine beyond its work capacity, degenerative changes, inflammations, senile alterations and disturbances in calcium metabolism.

Intervertebral disc calcifications may occur in the nucleus pulposus, the annulus fibrosus or in both areas together. Calcifications also appear in the cartilaginous end plates of the vertebral bodies, sometimes by extension from the annulus fibrosus. The most common location is the annulus fibrosus. The radiological differentiation of these sites can be identified with some degree of accuracy. Those calcifications situated in the middle of the intervertebral disc, and which are of uniform density, are more likely to be nuclear calcifications. These may be ovoid in configuration, and sometimes extend towards the posterior third of the intervertebral space. Calcifications in the annulus fibrosus are more frequent in the anterior, superior and inferior portions of the intervertebral disc. Occasionally calcium deposits occur in both areas simultaneously, and cannot be differentiated one from the other. Calcifications in the cartilaginous end plates alone are rather infrequent. No definite relationship with clinical complaints has been identified. Intervertebral discal calcifications in adults as a rule are symptomless, while those in children may be associated with painful syndromes. Adult intervertebral discal calcifications do not as a rule disappear, while those in children may or may not vanish. The reason for the occurrence of calcifications of the intervertebral discs in children, as well as for their disappearance and association with symptoms, is not known.

Calcification of the intervertebral discs in children is relatively infrequent. Brailsford mentioned that Barron (1926) had described discal calcification after trauma in a twelve-year-old boy. Brailsford himself noted that localized calcifications may occur in the intervertebral discs of children and be accompanied by backache, spasm, restricted motion and fever. In a report on discal calcifications in childhood Weens (1945) de-

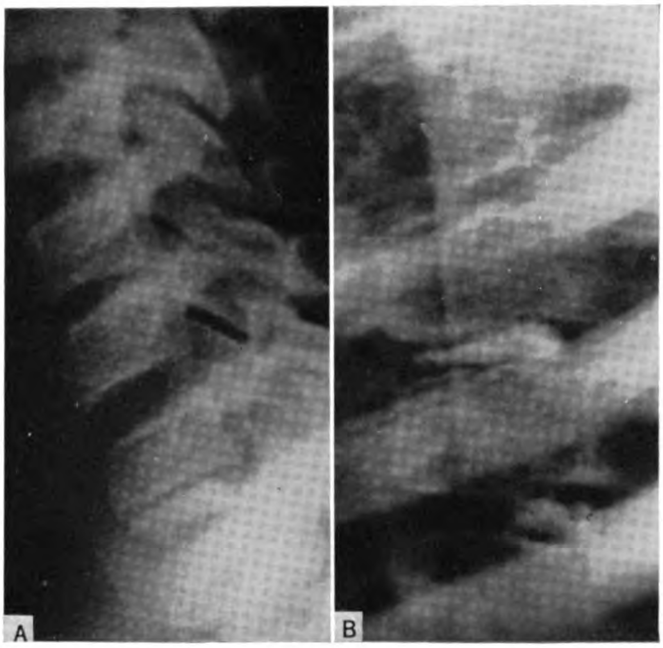

FIG. 284.—Calcification of nucleus pulposus between the sixth and seventh cervical vertebræ (*a*) and the fourth and fifth thoracic vertebræ and the fifth and sixth thoracic vertebræ (*b*). Severe pain was present in the lower neck and between the shoulder blades at the time these films were made. Subsequent examinations over a period of eighteen months showed no change in the discal calcifications even though pain had disappeared completely after several recurrences.

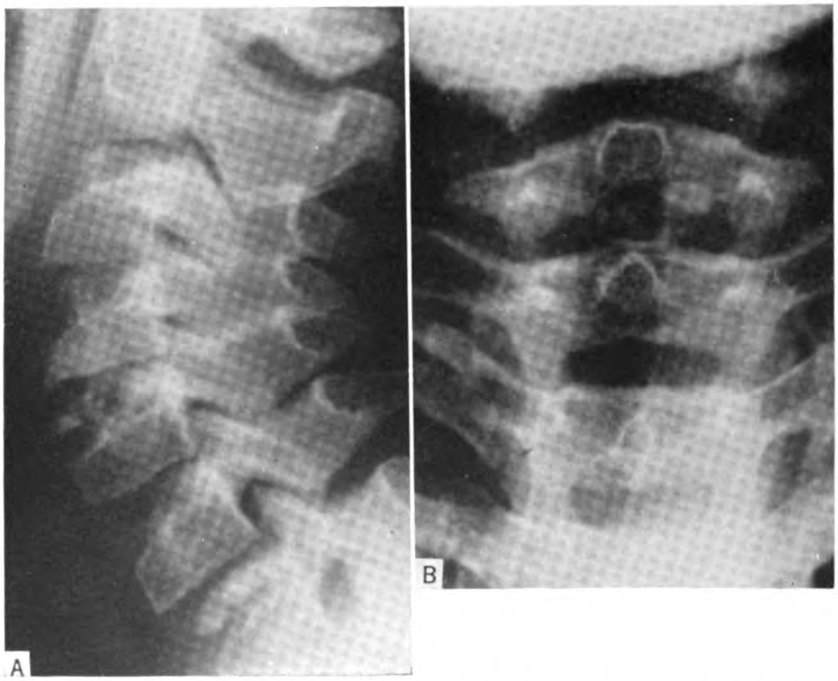

FIG. 285.—Calcifications between the fourth and fifth cervical vertebræ and the sixth and seventh cervical vertebræ in a four year old child with severe neck pain. There is an atypical wedging of the anterior aspect of the fourth cervical vertebra, and some increase in the density of the anterior two thirds of the body is present. (Courtesy of Dr. M. Boorstein, The Long Island Jewish Hospital).

scribed a five-year-old girl with pain in the lower cervical spine radiating to the back of her head. Roentgenograms revealed dense calcification in the interspace between the sixth and seventh cervical vertebræ, extending from the anterior to the midportion of the disc. Apparently these had been present for some time, inasmuch as a review of films made prior to hospitalization showed the lesion present then. Disappearance of the calcification was noted over a period of about four months. Weens gathered 5 cases of intervertebral disc calcification in childhood from the literature, and noted that pain was frequently present. There is a predilection for this condition to involve the lower cervical or upper thoracic spine at multiple levels. Of

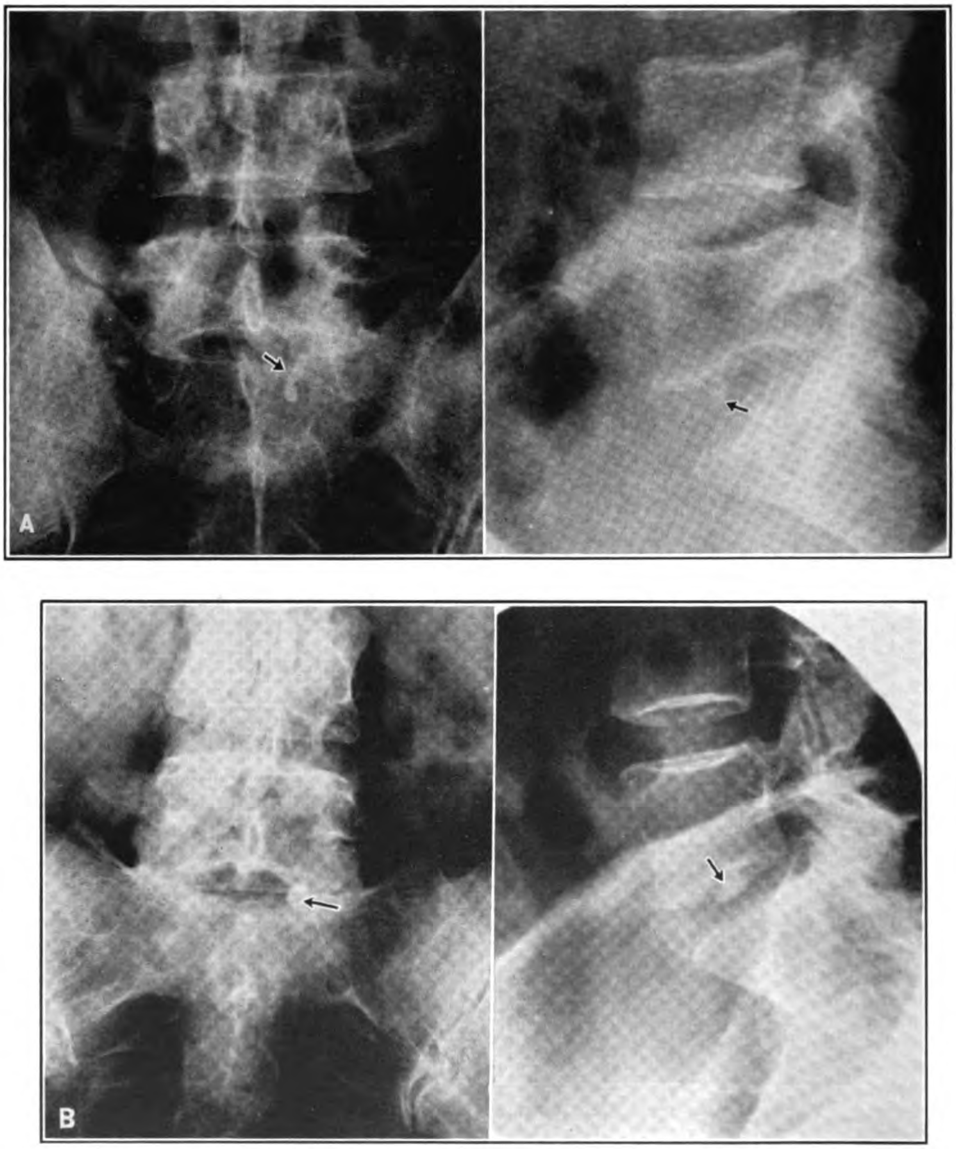

FIG. 286.—*A*, Calcification in the lateral aspect of the nucleus pulposus and adjoining annulus fibrosus at the lumbosacral interspace (arrows). *B*, Another case of local calcification of the annulus fibrous (arrows). The calcification is excentric, and is smooth.

particular interest is the fact that the calcifications have been known to disappear over a period of from several weeks to a few months or may persist for a period of years. Silverman (1954) added 7 cases to the 8 gathered from the literature, and stated that the condition was self-limited, requiring no treatment. He collected a series of 31 discs with calcifications in 15 patients. Eleven occurred in the cervical region, 17 in the thoracic spine and 3 in the upper lumbar spine. In two cases seen recently, persistence of the calcification in the cervical area was noted for a period of eighteen months, during which time pain on the neck disappeared and recurred several times in one (Fig. 284). The other (Fig. 285) had been seen because of neck pain and torticollis.

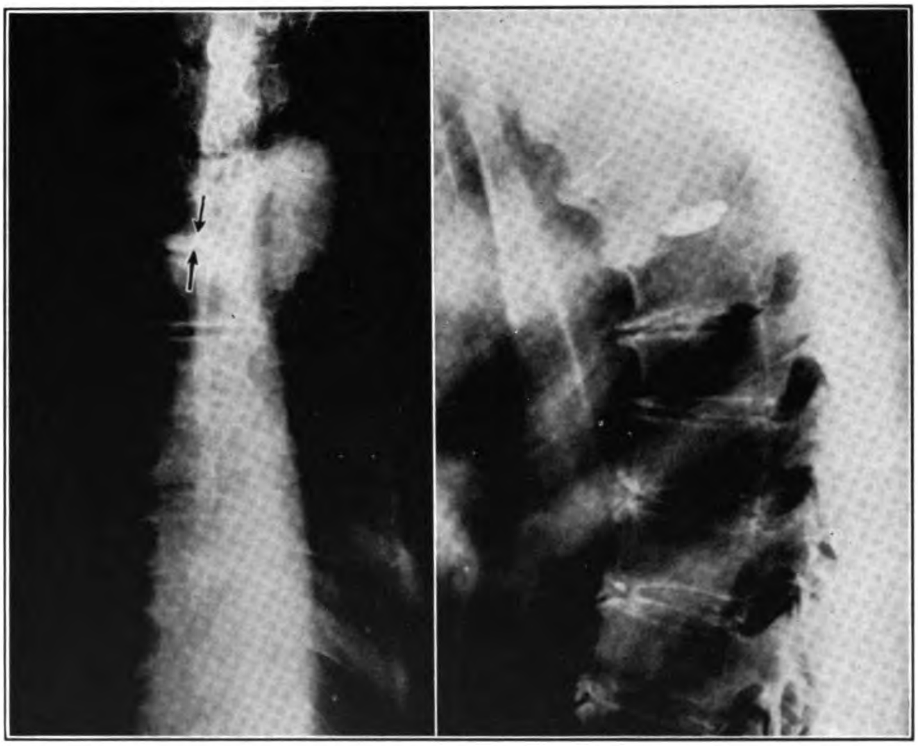

FIG. 287.—Symptomless calcification of the nucleus pulposus of the sixth thoracic intervertebral disc (arrows).

Sandström (1951) cited a case of a fifty-nine-year-old woman in whom calcifications in the first lumbar interspace disappeared. A second case concerned a fifty-nine-year-old male in whom calcifications were present in several of the lower thoracic intervertebral discs. Both had pain which was acute at the onset, with sensitivity to pressure and limitation of motion. A rapid sedimentation rate was present during the acute phase. Both these patients benefited from *x*-ray therapy. Sandström identified calcifications as occurring in the annulus fibrosus, usually of a permanent nature, and calcifications of the nucleus pulposus and the adjoining annulus fibrosus. The transitory variety is probably caused by an inflammatory process he termed myotendonitis calcarea, occurring in areas where tendons are at-

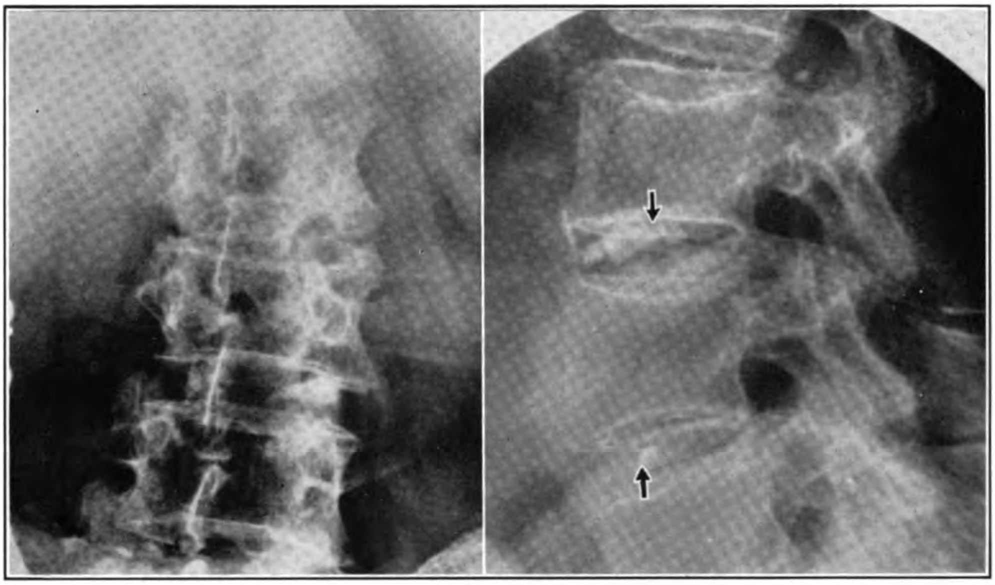

FIG. 288.—Multiple intervertebral discal calcifications (arrows), mainly of the annulus fibrosus.

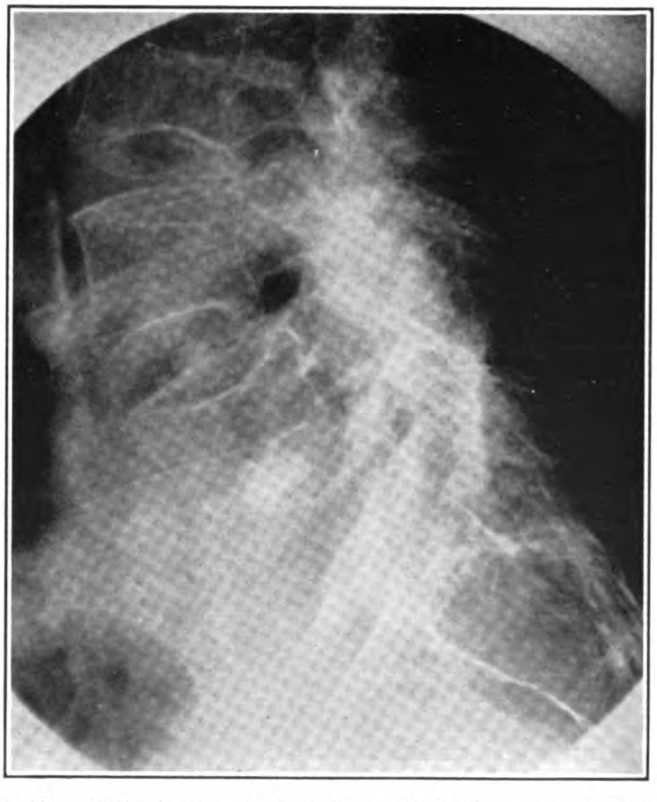

FIG. 289.—Multiple intervertebral discal calcifications in patient with myeloma and extensive osteoporosis.

tached to bone. The adjoining muscular tissue may also show such changes. When the disease is localized exclusively to tendinous tissues, such as the intervertebral disc, the name "tendonitis calcarea" is suggested by him. Sandström also mentioned that calcification induced by overdosage with vitamin D may be associated with impermanent intervertebral disc calcification.

Calcification of the annulus fibrosus, or less commonly, of the nucleus pulposus of a permanent nature is of no clinical significance. It occurs most frequently in the middorsal and lower lumbar regions, but instances of calcifications in the other parts of the spine are not uncommon. Calvé and Galland (1930) described a case of calcified intervertebral disc in which the entire nucleus pulposus had been converted into a mass of lime salt. Microscopically the calcium could not be seen, having been dissolved out during the preparation of the slide. In its place only strands of an amorphous residue could be observed. We have examined several cases of nucleus pulposus calcification at necropsy. In these cases all resiliency was lost, and the usual glistening appearance was replaced by a dull, grey appearance. The nucleus pulposus had been converted into a gritty, grey inelastic rubbery substance which merged imperceptibly with the surrounding tissue. The calcification was much more readily identified on the roentgenograms. With weakening of the posterior spinal ligament protrusion of a calcified

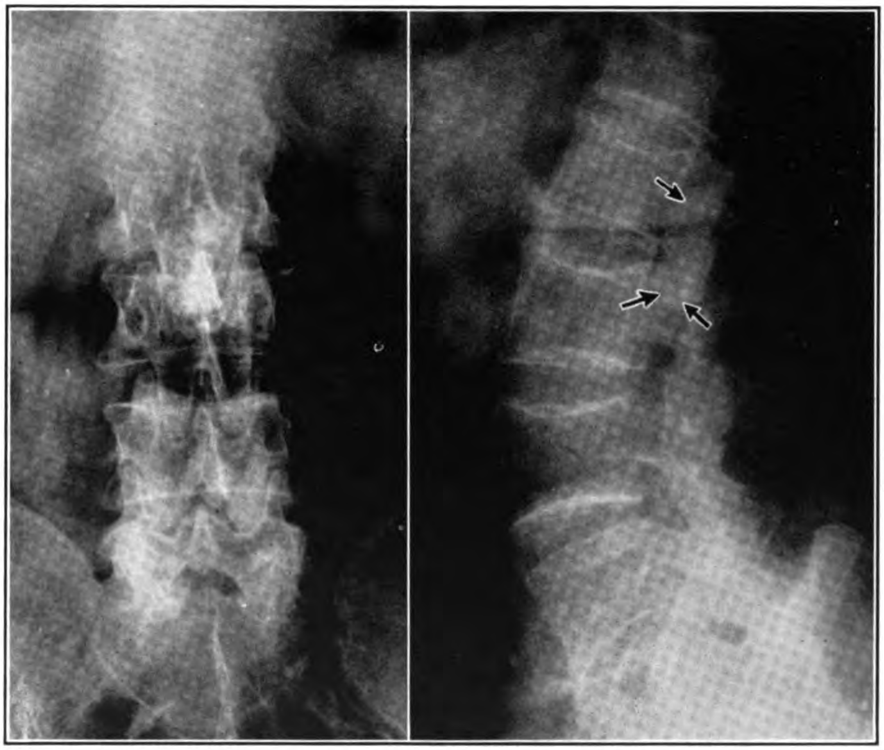

FIG. 290.—Calcified old extruded disc at the level of the second lumbar interspace (arrows) producing paraplegia.

nucleus pulposus may occur and severely damage the spinal cord. This is of special importance in the cervical and thoracic spine.

The occurrence of calcifications in the intervertebral discs of osteoporotic spines is not uncommon. We have seen this not only in the benign forms of osteoporosis, but also in multiple myeloma.

Rarely an intervertebral disc which has herniated into the spinal canal will further calcify. We saw one such patient (Fig. 290), a sixty-five-year-old woman who had a slowly progressing paraplegia attributed to a spinal cord tumor. Because the calcification within the spinal canal was quite apparent on plain films, it was assumed that she had a large spinal cord tumor, probably a meningioma. At operation a large, irregularly calcified mass identified as an old prolapsed disc was encountered. This was fixed firmly in position, and could not be dislodged. Only a decompressive laminectomy was done.

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"Vacuum Phenomenon" in Intervertebral Discs

Knuttson (1942) first called attention to the presence of radiolucent streaks at the lumbosacral intervertebral space, usually associated with a marked narrowing of the disc. He attributed this to desiccation of the disc in cases of spondylitis deformans. Inasmuch as the annulus fibrosus remains intact, irregular gas streaks corresponding to cracks in the dried portion of the disc appear. He mentioned that this was more apparent on roentgenograms made with the spine in extension, because widening of the intervertebral space increases the radiolucent area. The accentuation of the vacuum phenomenon in the hyperextended position was also described by Gershon-Cohen (1946). He too attributed this change to degeneration in the nucleus pulposus, and observed that no herniation of nuclear material into the vertebral bodies occurred when the vacuum phenomenon existed. Samuel (1948) attributed the presence of gas in the disc "vacuum" to diffusion from the blood.

In a survey of 2419 lumbosacral roentgenograms Marr (1953) found gas in the intervertebral discs in 2.026 per cent of cases, mostly in the lumbosacral interspace. This was associated with discal narrowing, marginal spurring and eburnation of the bony surfaces. He attributed the presence of gas to vaporization from surrounding fluids as a result of a partial vacuum presumed present in the intervertebral disc. He observed the radiolucent streaks mostly in the central portion of the disc. If the narrowing of the interspace was asymmetric, the gas seemed to collect in the narrowest portion. Raines (1953) attributed no clinical significance to this change. Gershon-Cohen, Schraer, Sklaroff and Blumberg (1954) observed that the vacuum phenomenon occurred also in multiple discs.

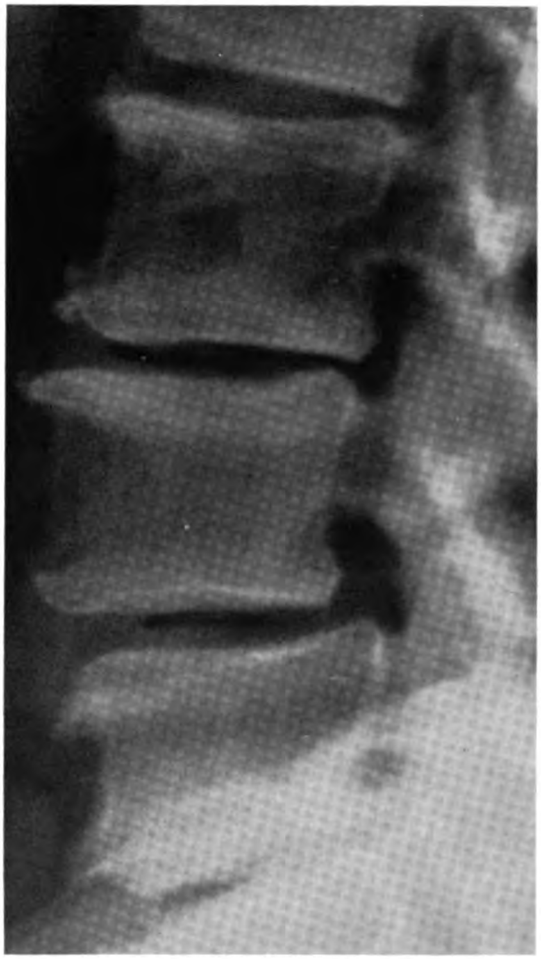

FIG. 291.—"Vacuum phenomenon" in the intervertebral discs between the third, fourth and fifth lumbar vertebræ and the first sacral vertebra. Incidental finding in a 59-year-old woman. Note slight anterior displacement of the fourth lumbar vertebra

The largest number of discs with the vacuum phenomenon in our series was three, usually the three lowermost lumbar interspaces (Fig. 291). A similar phenomenon is seen in the lower cervical spine occasionally, and somewhat more often in the small anterior spur formations present in the lumbar spine. Since the latter are due to intervertebral discal intrusions, a similar mechanism probably exists. The same change also has been observed in the symphysis pubis during and immediately after pregnancy. Occasionally difficulty is encountered in evaluating the linear gas streak because of overlying gaseous shadows in the bowel. Lateral body-section roentgenograms or stereoroentgenograms may be helpful in such cases.

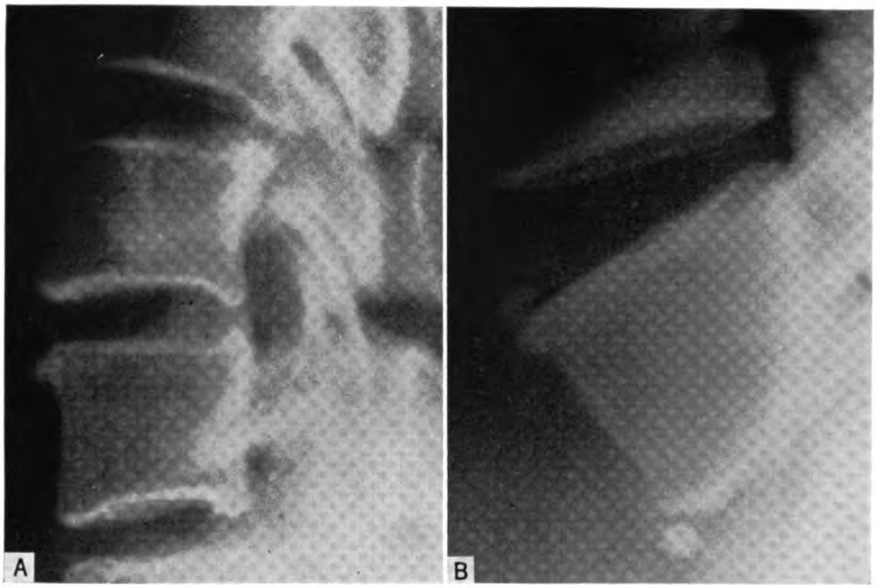

FIG. 292.—*A*, "Vacuum phenomenon" between fifth and sixth cervical vertebræ. Incidental findings in a 48-year-old man. *B*, "Vacuum phenomenon" at anterosuperior aspect of body of the fifth lumbar vertebra with fragment of bone separated by discal herniation. A similar change without the "vacuum phenomenon" is seen at the anteroinferior aspect of the fifth lumbar vertebra.

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HERNIATED DISCS

According to Mixter (1949) one of the earliest illustrations of a ruptured disc was that published by Charles Bell in 1824. This was described as a fracture, which undoubtedly was present. The illustration, reproduced in Mixter's paper, shows a fragment apparently partly disc and partly bone from the vertebra impinging against the spinal cord. Lindblom (1951) mentioned that there were drawings of ruptured intervertebral discs in two earlier texts, one by Weitbrecht in 1742, and the other by Henle in 1856. These had been made to illustrate the normal anatomy of the disc, and the pathologic nature of the lesion had not been recognized. Disc protrusion was described in 1857 by Virchow, who mentioned a cervical "ecchondrosis" with lethal compression of the spinal cord. In 1896, Kocher reported the case of a man who fell 100 feet, landing on his lower extremities. Death occurred because of rupture of the jejunum. Necropsy revealed protrusion of the intervertebral disc into the spinal canal at the level of the first lumbar vertebra, with no evidence of fracture. In 1911, Middleton and Teacher reported a case of injury to the spinal cord due to rupture of an intervertebral disc during muscular effort. This occurred in a man who felt a sudden sharp pain while lifting a metal plate. Paralysis soon developed and the patient expired a few hours thereafter. Necropsy revealed softening and hemorrhage about the lumbar enlargement in addition to a mass of cartilage lying in the spinal canal. Goldthwaite (1911) reported a patient who had sciatic pain followed by paraplegia after sacroiliac manipulation. Narrowing of the lumbosacral interspace was noted at operation, but no mention was made of exploration of the anterior epidural space. Mixter was of the opinion that this, too, was a ruptured disc. During the succeeding years there were isolated reports of injuries to the spinal cord attributed to enchondroma, which are now known to be herniated discs. Such reports were published by Frazier, Elsberg, Stookey, Ott and Adson, and Mixter (Mixter, 1949). Of particular interest is Dandy's report (1929) of 2 cases of transverse lesions of the cauda equina caused by free fragments of the intervertebral disc in the spinal canal. This report apparently was the first in which surgical treatment for ruptured discs was described, and the first to postulate that such fragments could cause sciatica. In 1930 Bucy collected 15 similar cases from the literature, and included 1 of his own. He regarded the lesions as chondromas of the intervertebral disc.

Recognition for attributing the syndrome of sciatica and low back pain to protrusions of the nucleus pulposus belongs to Mixter and Barr, who published their observations in 1934. Mixter's account of the events leading to the recognition of this syndrome should be read in the original (1949). In this communication he mentioned that Barr had read a paper on the same subject in 1933, but this had received such an indifferent reception that it was not published. Mixter mentioned the work of Mauric, published in 1933, which came to the same conclusions.

Lumbar Intervertebral Disc Herniations. Protrusions of the nucleus pulposus of the lumbar discs occurs most often in males in the third and fourth decades. The youngest patient with this lesion was reported by Fernström (1956) in an eleven-year-old girl. Love (1947) mentioned that of 1217 patients with herniated discs, 25 were between the ages of ten and nineteen years. Webb, Svien and Kennedy (1954) reported 5 patients fifteen years old or less at the time of operation. Herniated discs in patients beyond the fifth decade is by no means uncommon. The oldest patient in our series was well over seventy and the youngest was a fifteen-year-old boy.

The history of the patient is quite helpful. Not infrequently, particularly in a young active person, a history of trauma will be elicited. However, many patients, particularly females, have no history of obvious trauma. The type of pain associated with herniated disc varies with the extent and situation of the herniation as well as the patient's sensitivity to pain. Lateral and anterolateral displacements may compress the ipsilateral nerve roots producing unilateral sciatica. This is the most frequent complaint, occurring in about two-thirds to three-quarters of patients. Bilateral sciatica occurs in approximately 15 per cent, and may indicate a central protrusion with impingement against both sides. In the remaining individuals the distribution of pain may be atypical, shifting from side to side or involving only the low back. Pain may be exaggerated by straining, coughing or sneezing. Low back pain without sciatic radiation may be present with small mid-line discal protrusions. The onset of pain may be almost instantaneous, or gradual and progressive. In some cases the protrusion develops so rapidly that the acute compression of the cauda equina requires emergency care.

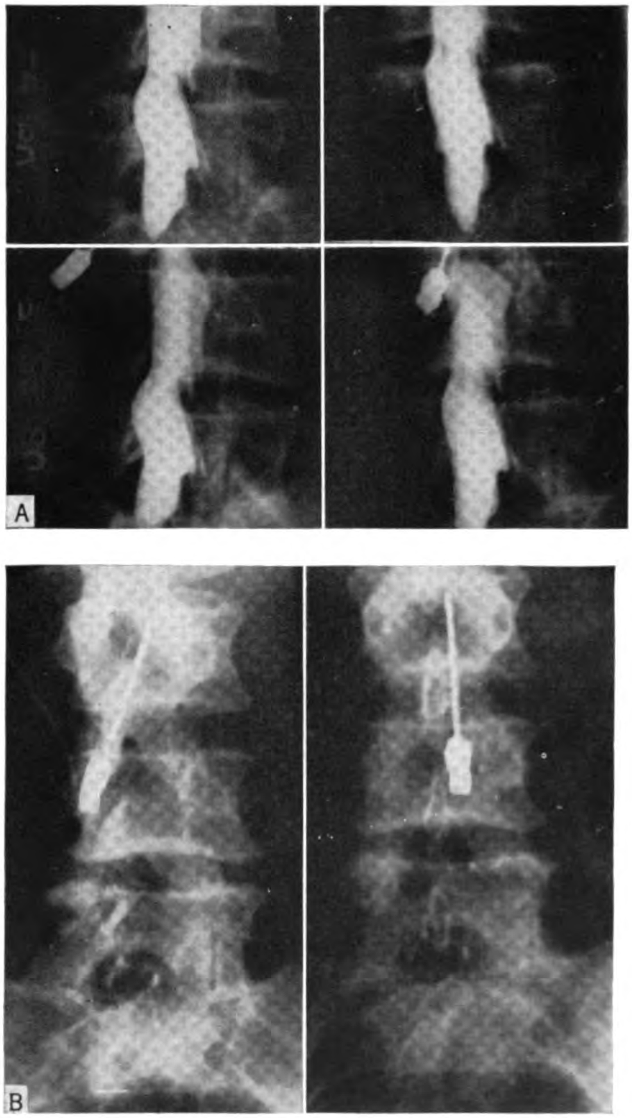

FIG. 293.—Myelogram showing small lateral defect at fourth lumbar interspace on the left side. Spot films with slight variations in angle of rotation of patient. Several droplets persist in the axillary pouches after removal of the pantopaque (*B*).

Physical examination may disclose loss of the usual lumbar lordotic curve, tilting of the spine away from the side of the lesion, tenderness over the spinous processes of the lower lumbar vertebræ, painful straight leg raising with the patient in the dorsal recumbent position, and tenderness on pressure over the course of the sciatic nerve. Diminished or absent

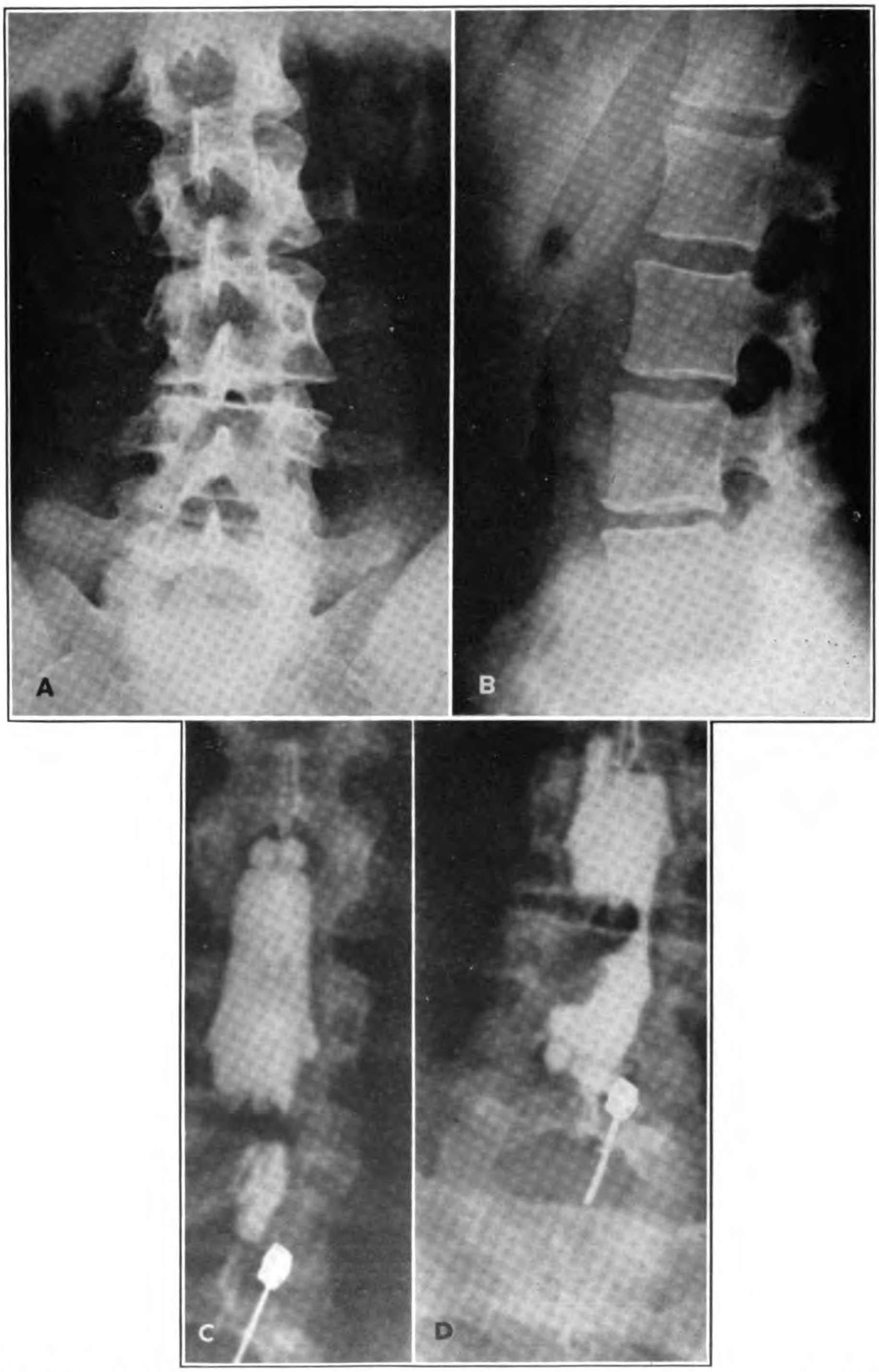

FIG. 294.—*A*, Narrowing of the third lumbar interspace and tilting of the spine to the right in a case of herniated disc. *B*, Same case, lateral view. The third lumbar intervertebral disc is narrowed, and the lumbar lordotic curve is straightened. *C*, Same case. Myelogram shows an extensive defect due to an extruded disc. The scalloped appearance of the lateral margin was due to rotation of the disc within the spinal canal. The herniation occurred at the third lumbar interspace.

ankle jerk or knee jerk reflexes, muscular weakness and atrophy of the limb on the involved side all may be present. Evaluation of the neurologic changes depends on understanding the peripheral nerve distribution. Thus, with discal protrusions involving the third lumbar interspace alteration changes in knee jerk reflexes may be encountered occasionally. Knee jerk reflex changes are uncommon in lesions involving the fourth or fifth lumbar interspaces. Diminution or absence of the ankle jerk reflex are common with herniated discs of the lumbosacral interspace, and are relatively infrequent at other levels. Areas of hypesthesia may be demonstrated depending on the level involved. Keegan (1943, 1944, 1947) presented dermatome charts for the trunk and extremities based on a study of the hypalgesic areas accompanying damage to a single nerve root. He pointed out that the neurologic deficits associated with lumbosacral disc herniations are easier to identify than those at the fourth lumbar interspace with compression of the fifth lumbar root. In the latter he found no reflex loss and the sensory changes were not easily found. McKenzie and Botterell (1942) concluded that 95 per cent of sciatica is associated with pressure on the roots of the first sacral or the fifth lumbar nerve. They were unable to distinguish between lesions of the fourth or fifth lumbar discs in 15 per cent of their cases.

Young (1947) considered neurologic signs of little help in the localization of herniated discs, and regarded such changes as only confirmatory in importance. Parrella and Zovickian (1950) pointed out that the neurologic changes seen with herniated discs were not constant. For example, 8 of their cases had hypesthesia of the fifth lumbar dermatome according to Keegan's chart. Of these, 3 had herniation of the fourth lumbar disc, 1 had a large disc protruding at the third lumbar interspace and the remaining 4 had fifth lumbar discal herniations. Absence or diminution of ankle jerk reflexes were encountered in 43 per cent of their cases. Sixteen had a lumbosacral protruded disc, and 4 had herniated discs at the fourth lumbar interspace. Davis, Martin and Goldstein (1952) reviewed the sensory pattern in 500 cases, and found no correlation between this and the level of the lesion. The extreme variability they encountered rendered dermatome charts unreliable.

Our own experience corresponds with the inconstancy of clinical localization observed by others. A review of 50 verified herniated discs at the fourth lumbar interspace revealed diminished ankle jerks on the side of the lesion in 17 cases. In 4 others, bilateral diminished ankle jerks were present, and this occurred when the lesion did not approach the opposite nerve root. Diminution of the knee jerk reflexes on the ipsilateral side was observed in 5 cases. No changes in the ankle or knee jerk reflexes were present in 24 instances. In 50 other cases of herniations at the lumbosacral interspace, knee jerk reflex changes were observed in 6 cases. Ankle jerk reflexes occurred in 35 others, and the remainder had no such changes. It becomes evident that while reflex alterations, when present, are of considerable diagnostic significance, they are neither constant nor absolutely reliable.

Patients with massive extrusions of the nucleus pulposus may present a syndrome of cauda equina compression indistinguishable from that caused by tumors. Symptoms of cord compression are accompanied by sciatic pain if the disc protrudes laterally as well as centrally. Ver Brugghen (1945) reported that out of 300 consecutive cases of herniated discs there were 9 with acute compression of the cauda equina by massive lumbar disc protrusions. These patients had saddle anesthesia, weakness of the legs below the knees, sphincter disturbances, and changes in the ankle reflexes in all the patients and knee jerk reflex changes in some. Eight were operated upon. In 3 the lesion occurred at the lumbosacral interspace, in 3 at the fourth lumbar interspace, and in 2 at the third lumbar interspace. French and Payne (1944) reported that cauda equina compression due to herniation of the nucleus pulposus was accompanied by bilateral sensory disturbances and reflex changes in 8 of their cases. The fact that massive extrusions of the disc may occur without the usual picture of cauda equina compression was brought out by Epstein (1949), who reported that of 150 lumbar myelograms there were 5 with complete spinal canal block. Only 1 of these patients had a syndrome suggestive of cauda equina compression.

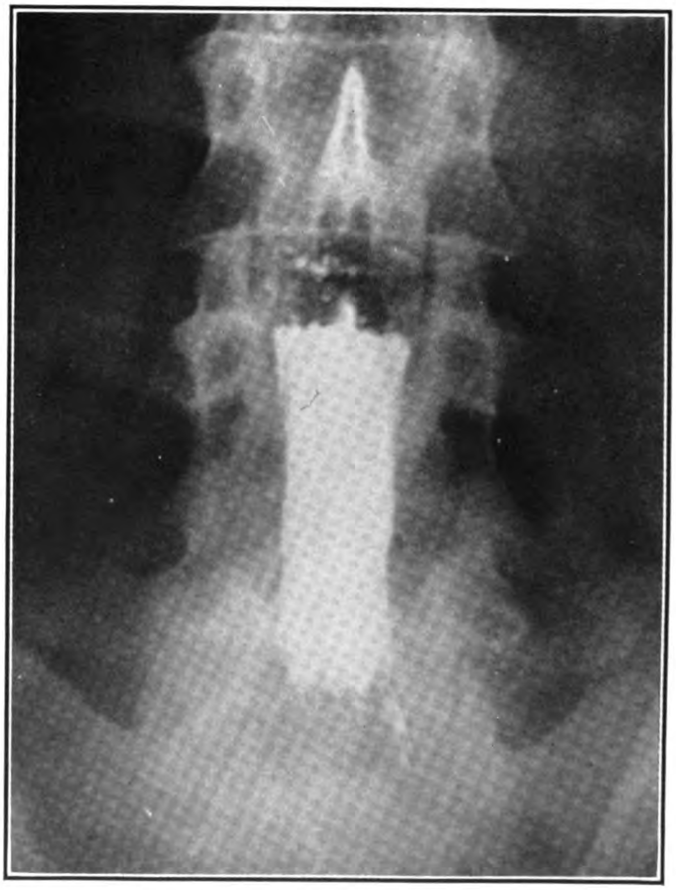

FIG. 295.—Complete block at the lumbosacral interspace due to a large herniated disc. The clinical picture in this thirty-eight-year-old woman was that of cauda equina pressure presumed due to a tumor. Complete relief of symptoms followed operation. Note the multiple dentate irregularities at the distal end of the pantopaque column.

The reasons for the wide variability of symptoms in patients with herniated discs are not entirely understood. Deucher and Love (1939) described edematous changes in protruded portions of intervertebral discs, and proposed that this may in part be responsible for the clinical recession of

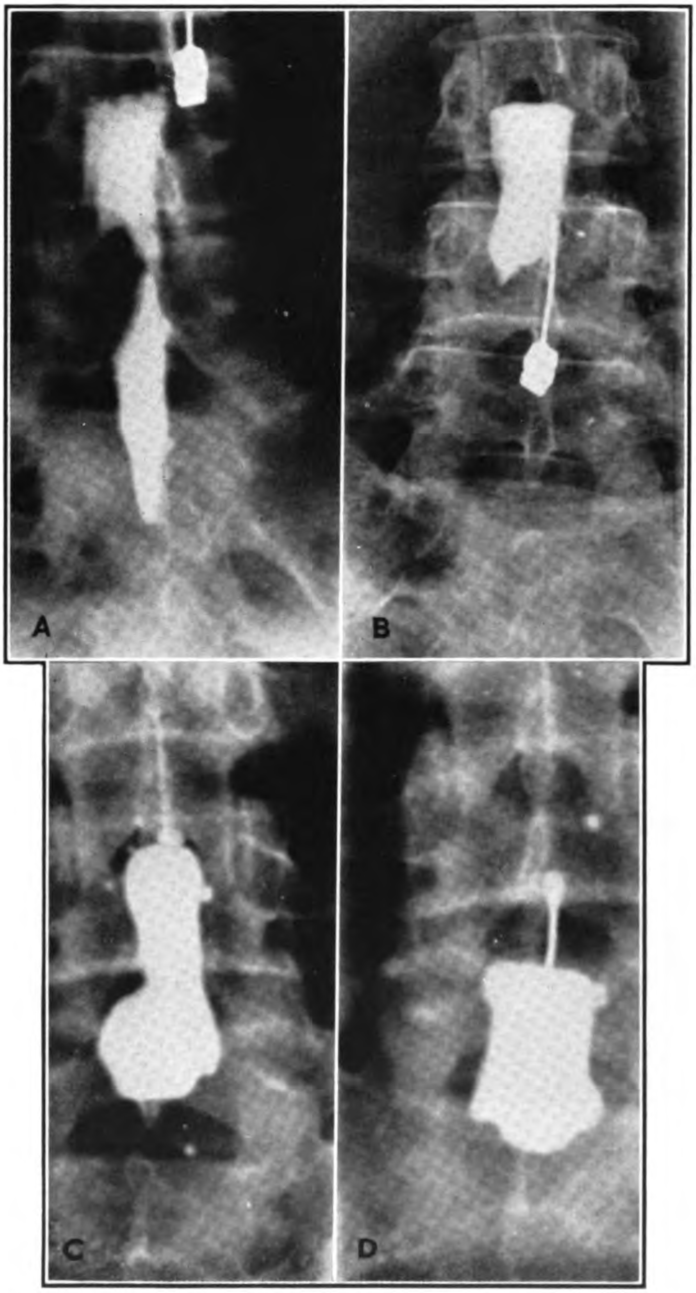

FIG. 296 —Three cases of massive distal herniation with intraspinal migration. *A*, Large extruded disc from the fifth lumbar interspace rotated in the spinal canal, producing an elongated defect extending well above the level of the discal defect. No defect is seen at the lumbosacral interspace. *B*, Extruded disc from the fourth lumbar interspace migrating cephalad, producing obstruction well above the involved interspace. *C* and *D*, Extruded disc from the third lumbar interspace migrating caudad, producing defect at the level of the fourth lumbar interspace.

symptoms. In a microscopic investigation of 100 removed discs they observed all parts normally present in unprotruded discs, but found that the tissues studied were altered by degenerative changes in cartilage cells, with a tendency for fibrous tissue to replace the normal fibrocartilaginous structures. Lindblom and Hultqvist (1950) suggested that protruded disc tissue might be absorbed. This was based on 160 necropsies, in which they observed all stages of change in the intervertebral disc, varying from radial rupture with beginning loss of tissue to almost complete disappearance of the nucleus pulposus and the annulus fibrosus. Absorption of prolapsed

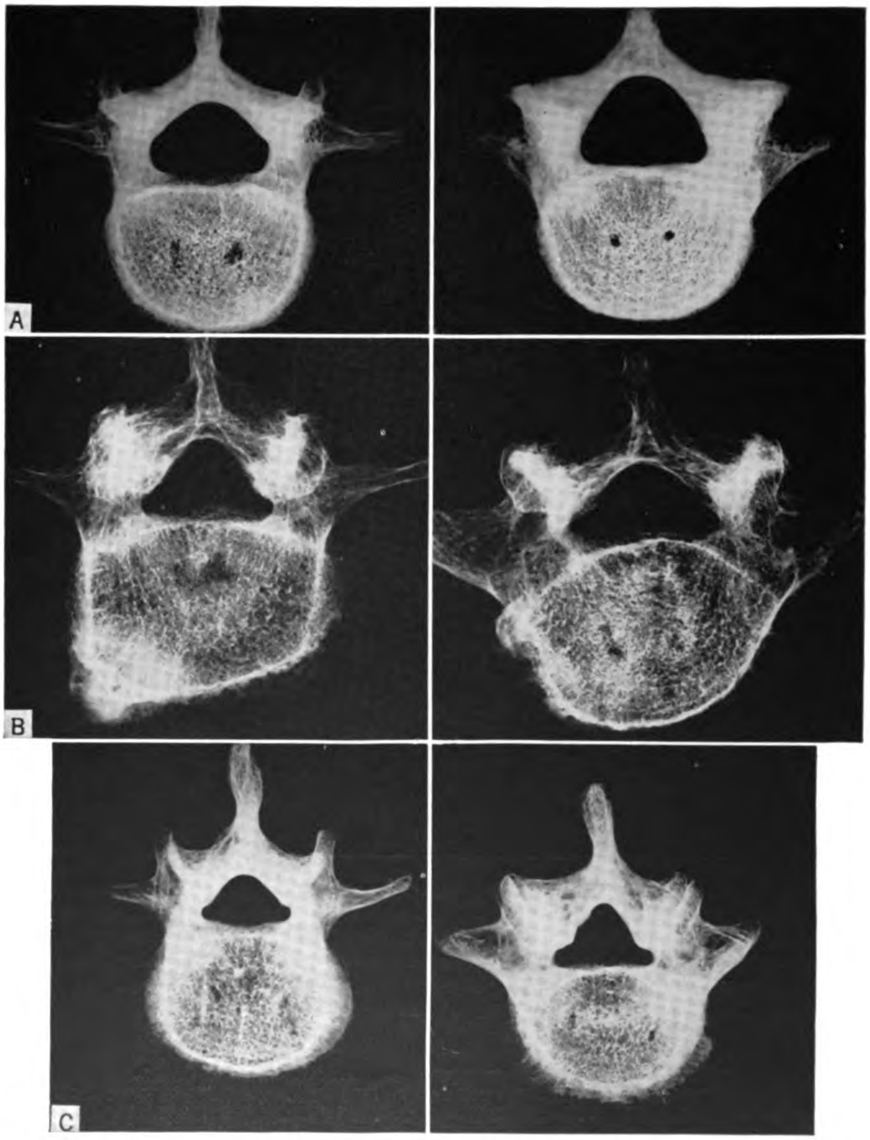

FIG. 297.—Roentgenograms of the fourth and fifth lumbar vertebræ to demonstrate configurations of the spinal foramina. *A*, Normal adolescent spine with wide lateral inferior angles. *B*, Adult specimen with extensive hypertrophic spondylosis. The lateral inferior angles are wide. *C*, Adult specimen with mild hypertrophic spondylosis. The angles are narrowed, more so in the fifth lumbar vertebra than in the fourth.

portions of an intervertebral disc may, in their opinion, result in the disappearance of symptoms.

Lateral, cephalad, caudad, intradural or subligamentous migrations of fragments of protruded disc material may occur and give rise to perplexing clinical pictures. Tucker (1956) mentioned two cases in which disc herniations at the first lumbar interspace entered the dural sac, causing a complete block. Kessler and Stern (1961) reported complete spinal canal block by posterior migration of a herniated lumbar disc. Greenwood, McGuire and Kimbell (1952) made special mention of a case in which herniated discal material from the third lumbar interspace had migrated downward to compress the fifth lumbar nerve root. Paterson and Gray (1952) reported that of 91 cases, 11 had fragments of disc tissue between the annulus fibrosus and the posterior spinal ligaments. In 8 others pieces of fibro-

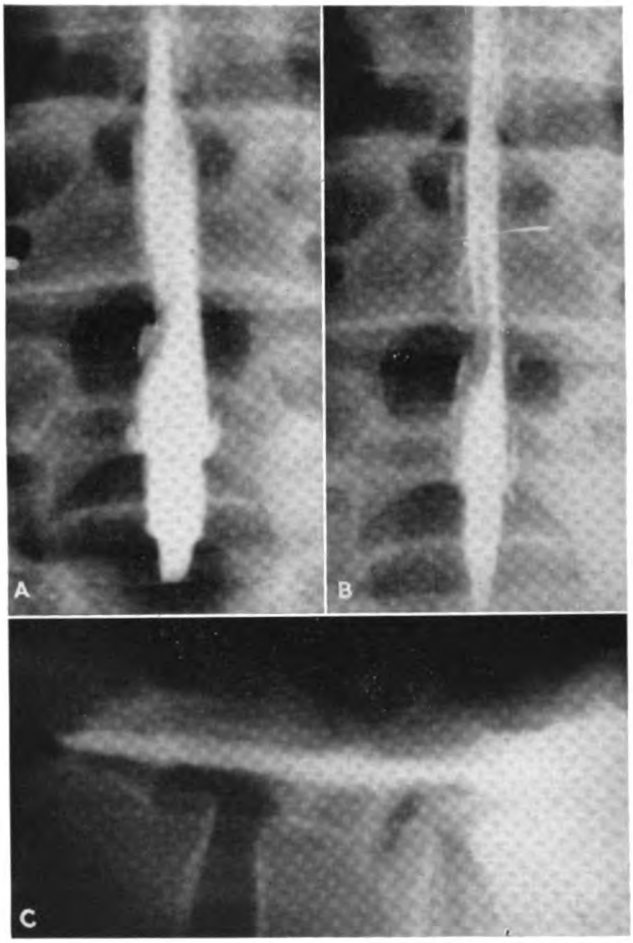

FIG. 298.—Herniated disc at fourth lumbar interspace on right side. A defect is present on the lateral aspect of the pantopaque column (*A*). The flow of pantopaque was slow, and thickening of a nerve root is seen (*B*). The lumbar puncture was quite difficult, and a free flow of cerebrospinal fluid had been obtained only at the second lumbar interspace. On the transabdominal film (*C*) it is noted that the thecal sac at the third and fourth lumbar levels is quite elevated. At operation a large herniated disc was found, and discal tissue had elevated the dural sac.

cartilage passed through the posterior spinal ligament and lay free in the spinal canal. In 2 still further displacements had occurred, the ventral being torn and the fragment entering within the thecal sac. Paterson and Gray mentioned that the clinical pictures in these cases were atypical, and were suggestive of a cauda equina tumor. Involvement of several nerve roots and of bladder function was common. If a hole is found in the annulus fibrosus at operation, and discal tissue is not readily observed and removed, further search may be required until the prolapsed material is discovered and removed. Even after removal, discal herniations can recur at the same interspace, (Fig. 299), or additional lesions may appear at other interspaces.

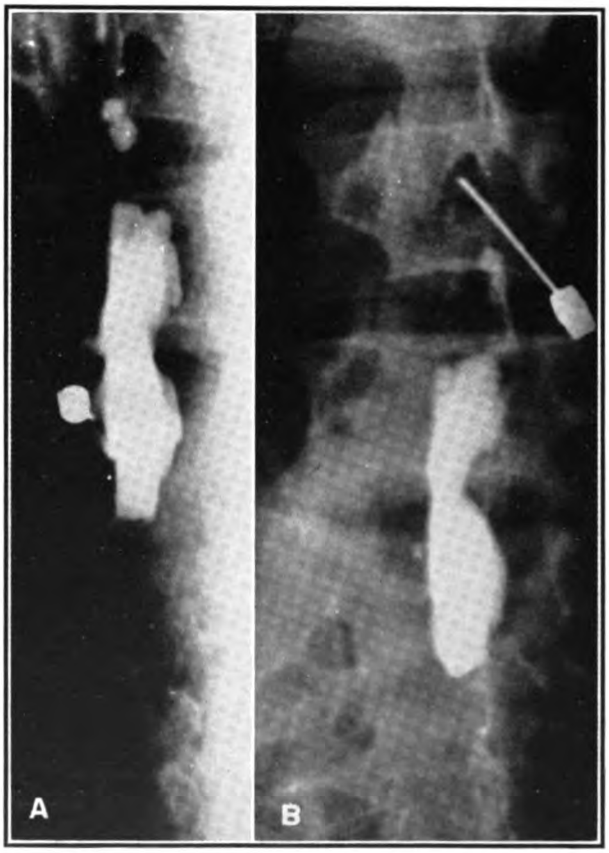

FIG. 299.—*A*, Lateral defect at the fifth lumbar interspace on the left side due to herniated disc, confirmed at operation. *B*, Same case, two years later. Symptoms had recurred, and operation disclosed further discal herniation.

It is possible for discal herniations to exist without causing clinical manifestations, provided that they do not intrude into the spinal canal or intervertebral foramina sufficiently to impinge significantly on the nerve roots or cauda equina. Not infrequently relatively large discal protrusions or spondylotic ridges produce myelographic defects unassociated with clinical manifestations. On the other hand, strategically situated spurs or small herniated discs can produce intense symptoms even though the myelographic defect is slight. One must keep in mind the concept of available space in the intervertebral foramina and the spinal canal, and realize that not only herniated discs but spondylotic spurs, bulging, softened but intact discs, thickening of the ligamenta flava, hypertrophy of the neural arches and variations in the sagittal diameter of spinal canals greatly influence pressure effects. Of great importance is the configuration of the lateral recesses of the spinal foramina, especially in the lowermost three lumbar vertebræ. The nerve trunk passing through the intervertebral foramen is in close proximity to the superior lip of the vertebral body and the inner aspect of the base of the pedicle. When this groove is constricted by thickening of the adjacent bone, the available space for the nerve root is diminished and the effect of small protrusions is enhanced. Unfortunately, the configuration of these lateral recesses cannot be estimated radiologically. It appears that narrowing of the sagittal diameter of the spinal canal to less than 1.5 cm. is associated with the possibility of constriction of the lateral recesses. In addition, narrowing of the canal exaggerates the pressure effects produced by other bony or ligamentous intrusions. The great sensitivity of the sciatic nerve to touch or pressure was verified by Smyth and Wright (1958). These workers experimentally pulled on the sciatic nerve after passing a suture around it at operation, controlling their observations by doing the same to the ligamentum flavum, the interspinous ligaments and the annulus fibrosus. They found that a simple pull on the thread sufficient to bring it in contact with the nerve root closely simulated the irritation and pressure of a herniate disc. The nerve need only to be touched to cause sciatica, and if touched repeatedly or continously the nerve root becomes hypersensitive. Dr. Joseph A. Epstein has repeatedly demonstrated at operation edematous and hemorrhagic nerve roots caught in constricted canals as described here when the discal herniation or osteophytic spur formation was disproportionately small in relation to the severity of the clinical syndrome. These patients were relieved only after vertical decompression of the narrowed lateral aspects of the canal was obtained surgically.

An investigation into the changes associated with degeneration of the intervertebral disc in a group of 100 spines of humans between forty and eighty years of age had been reported by Horowitz (1943). He mentioned that posterior bulging of the entire intervertebral disc without localized hernation was a common finding in adult cadavers. The posterior protrusions were formed not only by the dorsal bulge of the degenerated disc, but also by the hypertrophied posterior margins of the adjacent vertebræ. Softening of the intervertebral disc may be associated with narrowing of the space, together with a downward and backward settling of the upper vertebral body, which may be further exaggerated by bony overgrowth at the postero-inferior margin. Proliferation of bone on the inferior surfaces of the articular facets of the apophyseal joints also occurs. Such changes may be associated with myelographic defects indistinguishable from true protruded nucleus pulposus. In the lumbar spinal canal these may be asymptomatic because of the width of the available space, but in the cervical and thoracic regions pressure on the cord and nerve roots may be produced.

The anatomical effects of compression of the spinal nerves and ganglia by herniated discs were described by Lindblom and Rexed (1948), who removed specimens from 160 cadavers from fourteen to eighty-seven years of age. Among these were 60 instances of nerve compression, mostly caused by dorsolateral protrusion of intervertebral discs. Evidence was found of serious damage to the nervous structures, varying roughly with the degree of macroscopic compression. Forty-four of the damaged nerves and of the corresponding normal nerves were examined histologically by serial sectioning. The compression was shown to have produced serious deformity and atrophy of the spinal ganglion cells, fibrosis, and gross alterations of internal structure. Occasionally osteophytic enlargement of the intervertebral joint was an accessory factor in causing nerve damage.

Protrusion of the Intervertebral Disc on the Side Opposite Clinical Symptoms.—In 1949 Murphy reported 2 cases in which the clinical impression was that of a herniated disc on the left side, and myelograms disclosed right-sided protrusions. In both it was felt that if the exploration had been carried out only on the side in which symptoms indicated the lesion, the offending lesion would have been missed. Wycis (1950) reported 4 cases in which contralateral sciatic pain appeared after removal of a ruptured disc, the contralateral recurrence being at the same interspace. Three were verified at operation, and relief followed exploration of the contralateral side. He also reported a case in which ipsilateral symptoms were relieved by contralateral removal of a partly herniated disc. Wycis believed that bilateral exploration might be required in view of the fact that a nucleus pulposus may be herniated bilaterally, resulting in a dumb-bell deformity. Kaplan (1949) also reported 2 cases in which herniated discs were associated with signs and symptoms on the opposite side. We, too, have observed 3 cases in which exploration on the side of long-standing pain did not disclose the lesion, while myelography correctly indicated its location.

Atypical myelographic defects may be due to migration of herniated nuclear material (Figs. 294, 295). In the presence of a mid-line discal herniation, symptoms may vary from side to side. Due precaution must be exercised in evaluating clinical as well as myelographic observations to make sure that the proper, or both sides, are explored when necessary. In this regard, it is well to recall that with mid-line large protrusions, the lesion may be missed if only the lateral aspects of the spinal canal are explored, and a transdural exploration may be required.

Plain Film Roentgenograms in the Diagnosis of Lumbar Herniated Discs.—Plain film roentgenograms of the spine should routinely precede myelography. Their importance lies more in the elimination of other causes of back and radicular pain than as an indication of disc herniation. In a review of 300 verified cases of herniated disc at the lumbosacral interspace, the plain film roentgenograms were normal in 64 per cent of the cases. The lumbosacral interspace was markedly narrowed in 16 per cent, and in the remaining 20 per cent slight to moderate narrowing was seen. In 12 per cent of the cases narrowing of the fourth lumbar interspace was observed. Osteophytic reaction at the lumbosacral interspace was present in about one-third of our cases, and in most the extent of the change was considered within normal limits for the age groups. Lateral films revealed straightening of the lordotic curve in one-third of the patients. Tilting of the spine towards the side of the lesion was relatively infrequent, being seen in 10 per cent of cases. In occasional instances the lumbar spine was tilted away from the side of the lesion.

Data were available in 50 cases in which more elaborate plain film studies had been made. Films were taken in flexion and extension in the erect lateral posture. Examinations were also made in the anteroposterior position with the patient bent sharply to the right and left. These examinations were not helpful in identifying either the presence or location of a herniated disc. In more than half the cases changes at the articular facets of the lumbosacral joint were seen. The facets were either asymmetric or coronally placed. It was not believed that this was of diagnostic help, inasmuch as similar changes were observed regularly in patients without any back complaint. This concept is at variance with that expressed by Gillespie (1949), who reviewed 500 normal patients and 500 others in whom laminectomy had been done for disc protrusions. He found that 34.2 per cent of the laminectomy series showed congenital spinal instability as compared with 8.8 per cent of the control series. On the other hand, Peyton and Simmons (1947) and Splitthoff (1953) noted no significant roentgenographic difference in the lumbosacral junction of patients with and without backache. Gage and Shafer (1953) also concluded that plain films were unsatisfactory in the diagnosis of herniation of the lumbar intervertebral discs. Armstrong and his co-workers (1950) asserted that disc lesions could not be attributed to congenital defects of the disc proper, and that abnormalities in the planes of the articular facets were not an important cause of low back pain. A narrowed lumbosacral interspace with sacralization of the fifth lumbar vertebra was of no diagnostic significance.

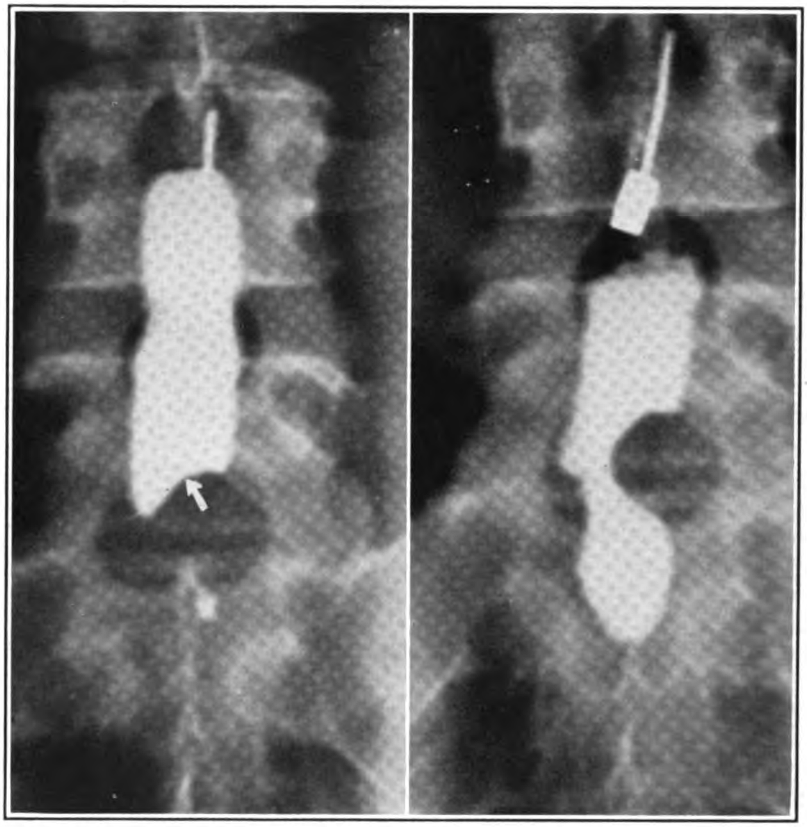

FIG. 300.—*A*, Spot roentgenogram made as head of pantopaque column approached the lumbosacral interspace while patient was being moved towards erect position. Note the arcuate appearance of the pantopaque (arrows). *B*, Same case, a few seconds later. The pantopaque column is deformed both on the right and left sides, producing a dumb-bell deformity, more marked on the left. Bilateral discal herniation.

Herniation of the Fourth Lumbar Intervertebral Disc.—The plain film roentgenograms may be somewhat more helpful in the identification of discal protrusions at the fourth lumbar interspace. In a review of 125 consecutive cases in our series, the plain films were normal in 55 per cent In 25 per cent there was a significant narrowing at the fourth lumbar interspace. In the remaining cases a slight narrowing was present, but this was not sufficient to warrant any definite conclusions. There were several disconcerting observations in this series. In 18 cases there was marked

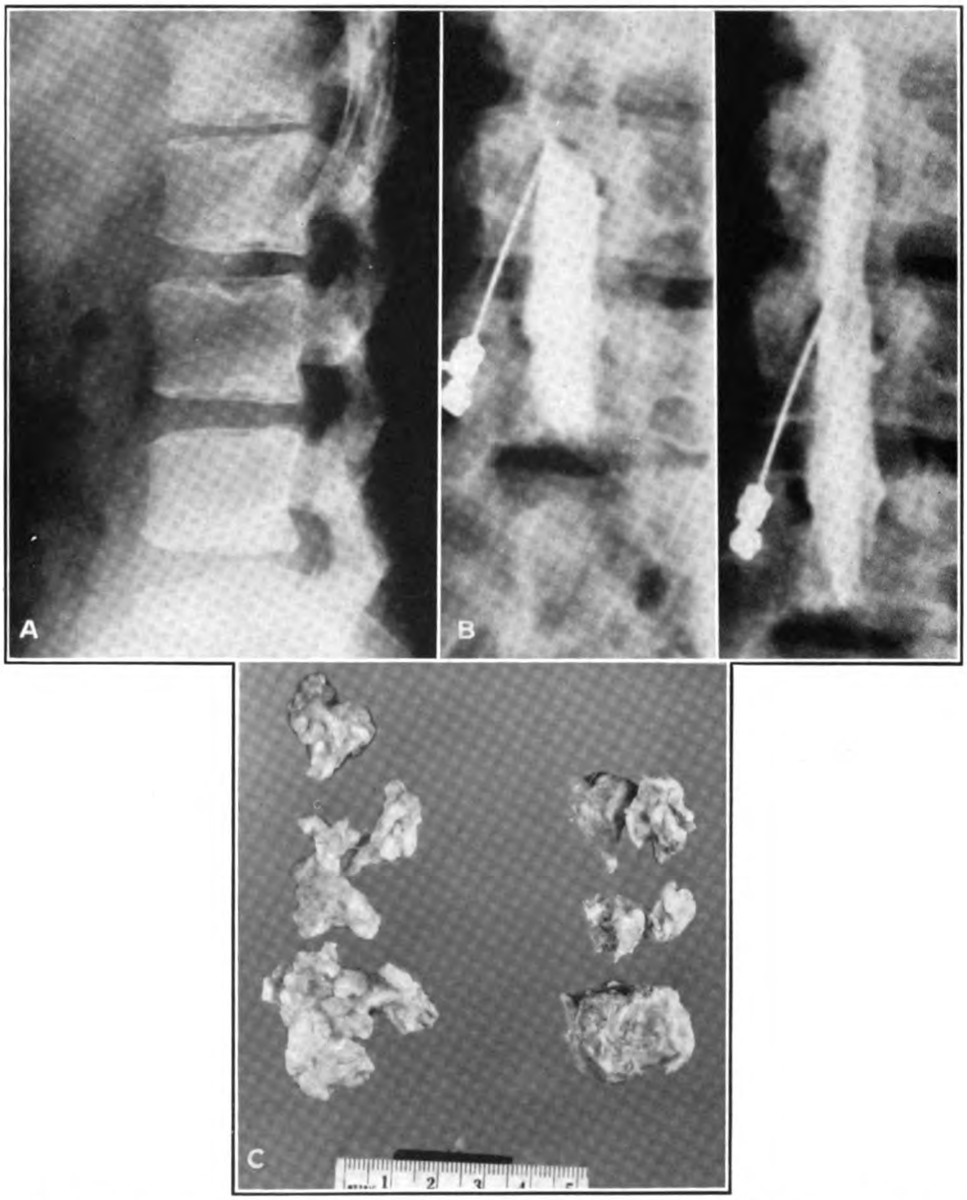

FIG. 301.—*A*, The lumbar lordotic curve is straightened, and multiple Schmorl's nodes are present. *B*, Same case, myelograms reveal complete block at the lumbosacral interspace. Note linear streaking within the pantopaque column due to edema of the roots of the cauda equina. *C*, Same case, operative specimen.

narrowing at the lumbosacral interspace, and in 10 of these patients, all over forty-five years of age, significant arthritic changes were present. Of this group, 3 also had a slight narrowing of the fourth lumbar interspace. In 4 others, narrowing of the third lumbar interspace was seen.

Straightening of the lumbar lordotic curve was oberved in one-fourth of this series, and when present, was presumed to be of some diagnostic help. The absence of a straightened lumbar lordotic curve was not considered significant. In 4 cases the lumbar spine was tilted contralateral to the affected side, and in 2 the spine was inclined towards the affected side.

In 12 cases the narrowing of the fourth lumbar interspace as observed in anteroposterior views was irregular. In 7 of these patients the narrowing was more marked on the side towards which the nucleus pulposus was herniated. In 4 instances there was a shifting of the fourth on the fifth lumbar vertebra laterally. In all these the shift was towards the side of symptoms. On reviewing the operative findings no relationship could be established between the size of the herniated disc and the degree of change noted on the plain films.

Roentgenography of the articular facets, the intervertebral foramina and films made in flexion, extension, and bending to the right and left were not helpful in the diagnosis of fourth lumbar herniated discs.

Herniated Discs in the Upper Lumbar Spine.—There were 10 such cases in our series, 2 at the first lumbar interspace, 3 at the second and 5 at the third lumbar interspace. The plain films in all except one (Fig. 294) were normal. Peyton and Simmons (1947) indicated that routine roentgenograms were not helpful. They noted that of 90 patients examined, 16 had narrowing of the corresponding interspace. However, similar narrowing was seen at other levels as well. In a discussion of a paper by Graf and Hamby (1953) Beswick incidentally mentioned a case which had a striking narrowing of one interspace. He felt this to be a reasonable localization without myelographic confirmation. Myelography, however, showed the lesion to lie two interspaces higher, and this was confirmed at operation.

Myelographic Diagnosis of Herniated Lumbar Discs.—Although an accurate diagnosis of a herniated disc can be, in the opinion of many, reasonably established on the clinical aspects alone, myelography should not be disregarded. When capably performed, myelography is a relatively painless and safe procedure. The ease and skill with which it is performed is the chief factor in the patient's response, and myelography should not be relegated to anyone untrained or unskilled. The procedure requires understanding and delicacy in manipulation. Even in the most experienced hands, considerable distress may be caused the patient. Nevertheless, in the same way and for the same reasons that colon roentgenograms, for instance, are made before operation for a bowel lesion, so should myelography be done when surgery of the spinal canal is contemplated. Indeed, in my opinion an exploration of the spinal canal without preliminary myelography even when the clinical picture is presumably complete, is comparable to laparotomy without previous adequate investigation. To carry this a bit further, even at the risk of being tedious, there are instances in which *x*-ray examinations fail to reveal an intra-abdominal lesion, and laparotomy is done nevertheless. Similarly, myelography may fail to disclose a definite lesion, and still surgical intervention is required because of the over-all picture. In either case operation is not contraindicated by a presumably negative *x*-ray study.

It should be routine to investigate the spinal canal well above the site of the suspected lesion. The upper level for lumbar myelography should be at the middorsal spine, and especially if the lumbar area is normal, the cervical region should be checked as well. I have in mind several instances in which symptoms referable to the lower extremities were caused by lesions in the cervical spinal canal, and which had been misdiagnosed because cervical myelography was not done.

Unless fluoroscopy is carefully performed, there is a possibility of overlooking a small defect because of overdistention of the thecal sac or of missing a lateral or midline small defect. Recently I saw a patient in whom

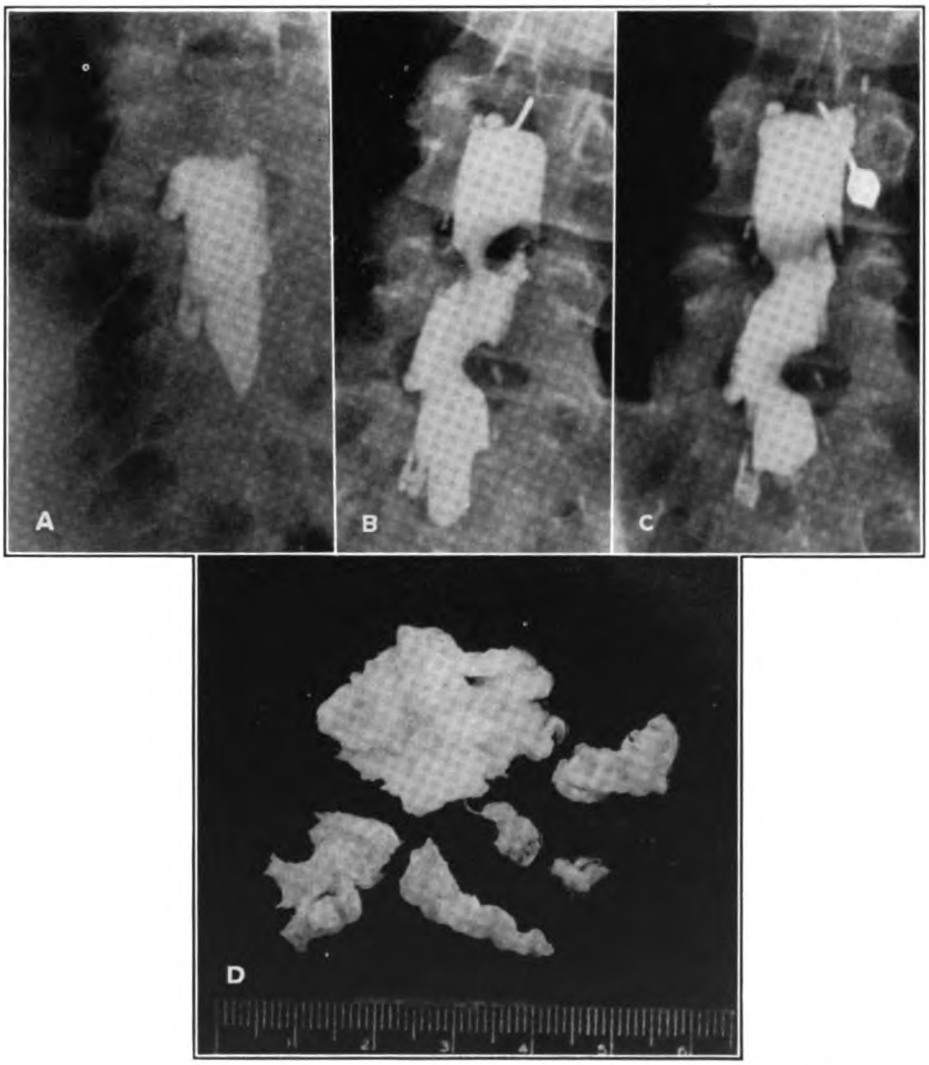

FIG. 302.—*A*, Erect myelogram with 10 cc. of pantopaque. The right axillary pouch is large, that on the left side is not visible, but no defect in the column itself is present. *B*, Film made while patient was being turned downwards. Note the dumb-bell defect at the fourth lumbar interspace and the left lateral defect at the fifth lumbar interspace. *C*, Same case, a few seconds later. The defect at the fourth lumbar interspace has assumed a veil-like appearance, while that at the lumbosacral interspace is unchanged. *D*, Same case. Operative specimens. The upper one is from the fourth lumbar interspace, the lower from the lumbosacral interspace.

a definite protrusion was identified at the fourth lumbar interspace. On following the column down, a defect was also found at the lumbosacral interspace. This was done using 4 c.cm. of pantopaque. The study was continued after an additional 6 c.cm. of pantopaque was added, so that a total of 10 c.cm. were used. This time the defect at the fourth lumbar interspace was again identified, but the caudal sac was so distended that the lumbosacral defect was obliterated when the patient was erect. However, this defect reappeared when the patient was tilted downwards. Operation revealed herniated discs at both the fourth and fifth lumbar interspaces. It has been pointed out by others, and repeatedly seen by us, that small defects are detected most readily fluoroscopically and by spot film roentgenograms made at the time when a lateral or veil defect appears in the pantopaque column. Such lesions may be obliterated by overfilling the thecal sac, and the great importance of watching the ends of the pantopaque column in oblique and lateral as well as frontal positions during its flow is re-emphasized.

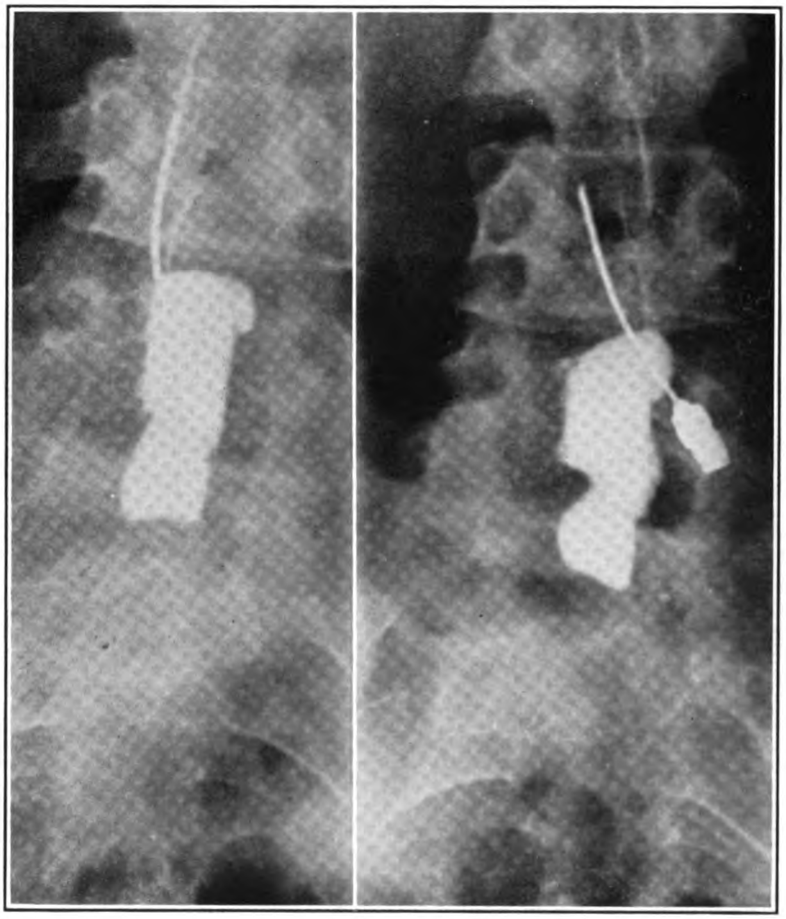

FIG. 303.—*A*, Myelogram showing slight indentation and veil-like appearance of the pantopaque column at the lumbosacral interspace. *B*, Slight rotation towards the right brings out the left lateral defect to better advantage.

The illustrations presented here depict some of the many filling defects seen with herniated discs. The absence of a filling defect does not in itself rule out a herniated disc, especially when the thecal sac is relatively narrow or high and the spinal canal is wide. In such instances a herniated disc may not impinge against the dura and the myelogram appears normal. Careful review of the clinical aspects may lead to surgical intervention nevertheless.

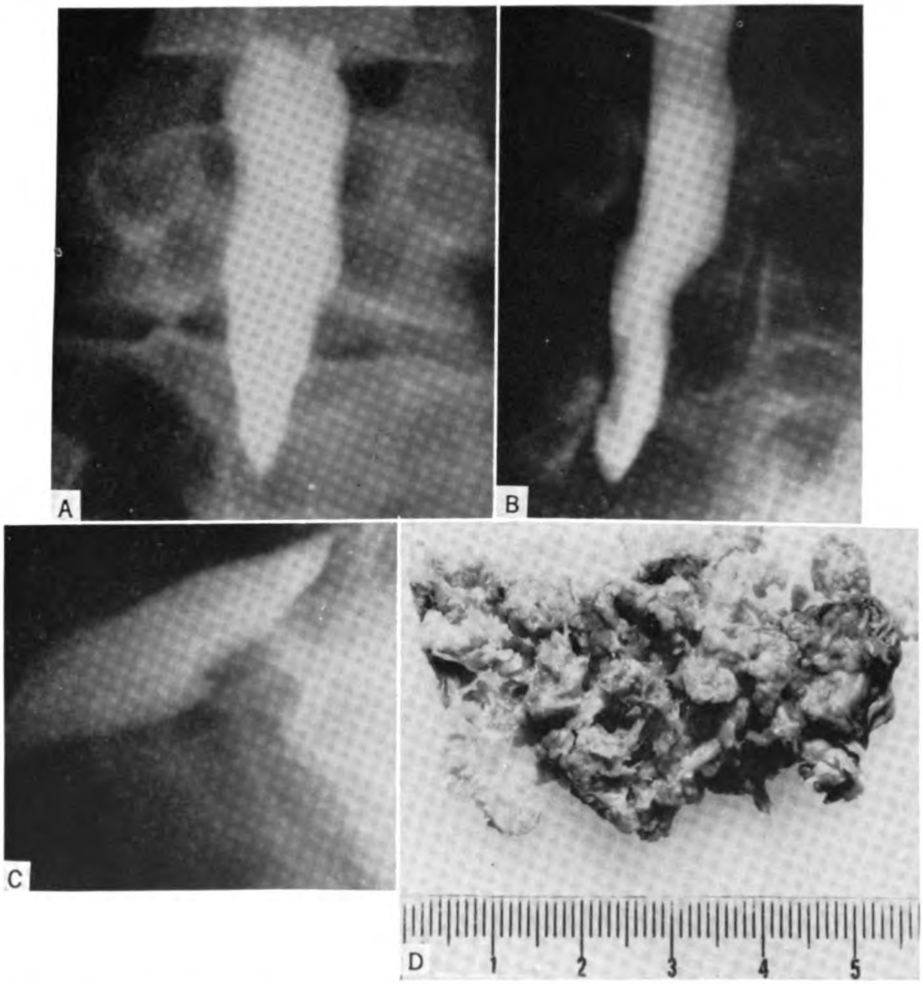

FIG. 204.—The apparent size of a myelographic defect is not necessarily indicative of the size of the discal herniation. In this patient, a 39-year-old woman with an extruded disc, the defect was minimal on the anteroposterior film (*A*), seen better on an oblique view (*B*), and the caudal sac was elevated as seen on the transabdominal view (*C*). The surgical specimen is shown in (*D*).

Depending on the location and extent of a lesion, four major varieties of filling defects appear. When the protruded disc lies laterally, the defect is merely an obliteration of the corresponding axillary pouch. This may be so slight that the pantopaque column remains unindented. In most instances, however, a slight convexity of the opaque column is co-existent with obliteration or elevation of the axillary pouch. More obvious lesions

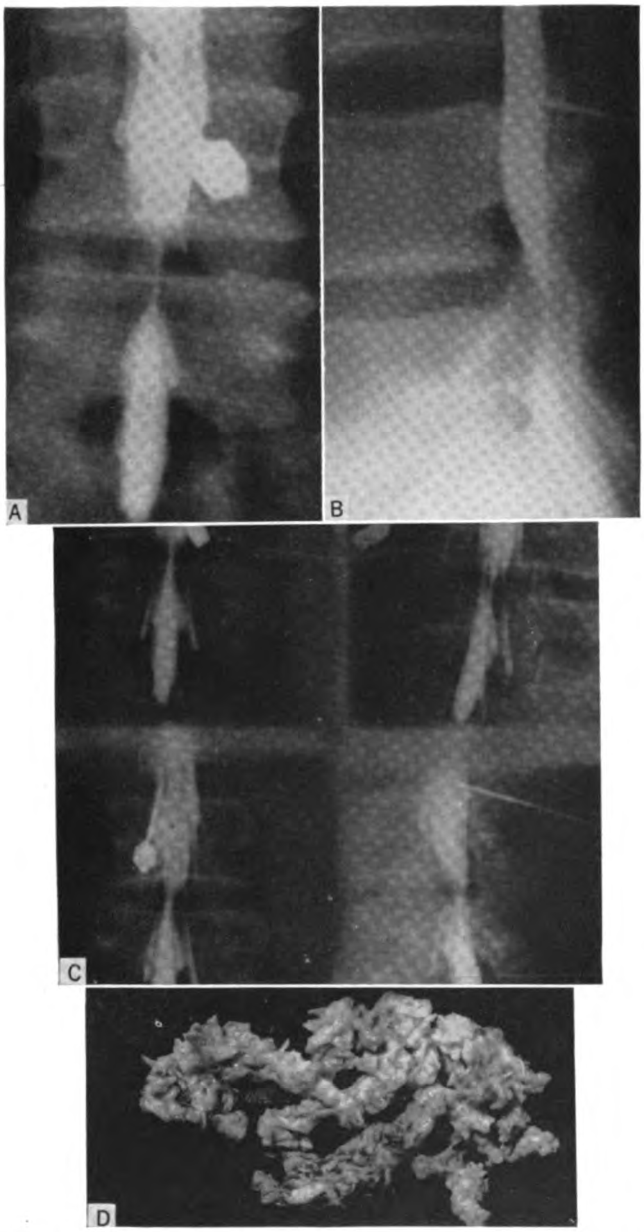

Fig. 305.—Large extruded disc at fourth lumbar interspace. In the anteroposterior view a thin channel is seen in the midline (*A*). As seen on the transabdominal view the herniated disc projects upwards, displacing the pantopaque towards the upper aspect of the canal (*B*). Multiple spot films during fluoroscopic inspection show the defect on the right and left sides, and thickened nerve roots displaced medially (*C*). The operative specimen (*D*) is photographed approximately 1:1.

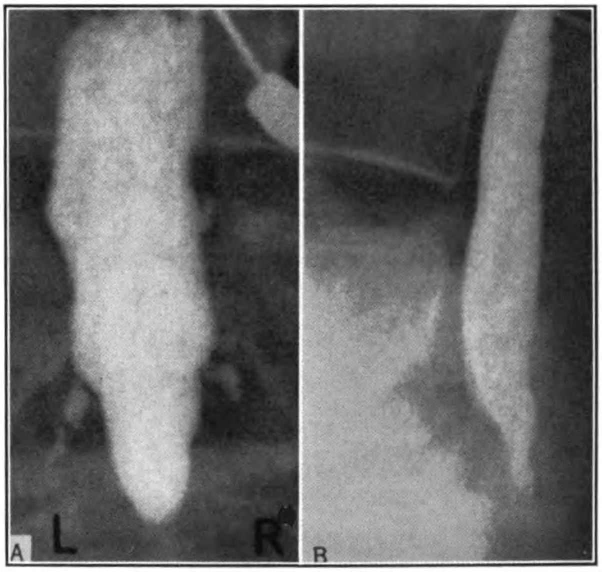

FIG. 306.—Midline herniated disc in a thirty-three-year old woman with low back pain in the midline radiating down both legs, more marked on the right side. The anteroposterior myelogram (*A*) shows a minimal elevation of the right axillary pouch. The transabdominal film (*B*) indicates the presence of a large midline herniation.

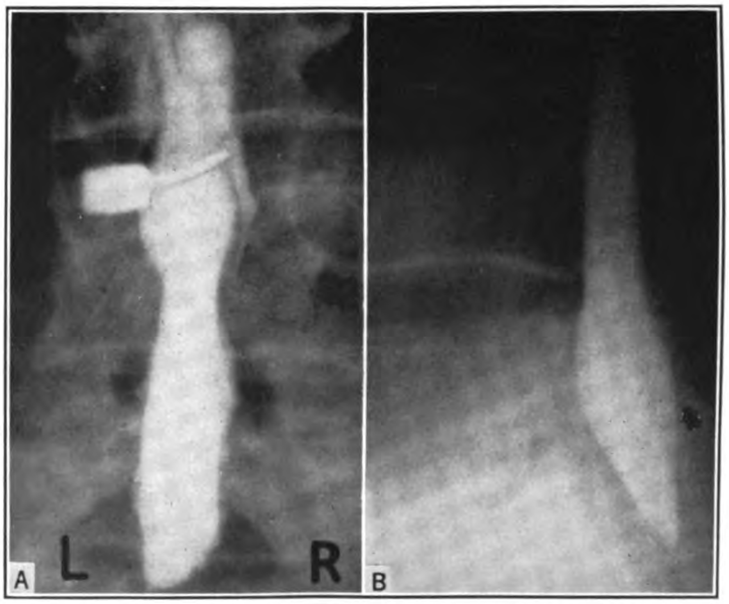

FIG. 307.—Lateral defect on the right side of the pantopaque column at the level of the body of the fourth lumbar vertebra (*A*). The transabdominal study (*B*) shows no elevation of the column. At operation the defect was proved to be due to a herniated disc which had extruded from the fourth lumbar interspace and moved upwards to a parallel position in the spinal canal. Note the medial deflection of one of the lateral nerve roots by the needle tip at the third lumbar interspace and parallel to the filling defect as well.

are manifested by a greater indentation, varying from smooth shallow defects to elongated, oval or semicircular impressions which may be scalloped. In these cases the axillary pouch usually is obliterated, particularly with the larger lesions. The elongated defects are observed when an extruded disc rotates upon itself within the spinal canal and comes to lie parallel to the thecal sac. In some the deformity extends across the midline and indents the contralateral side, particularly when nuclear extrusion is through multiple breaks in the posterior spinal ligament.

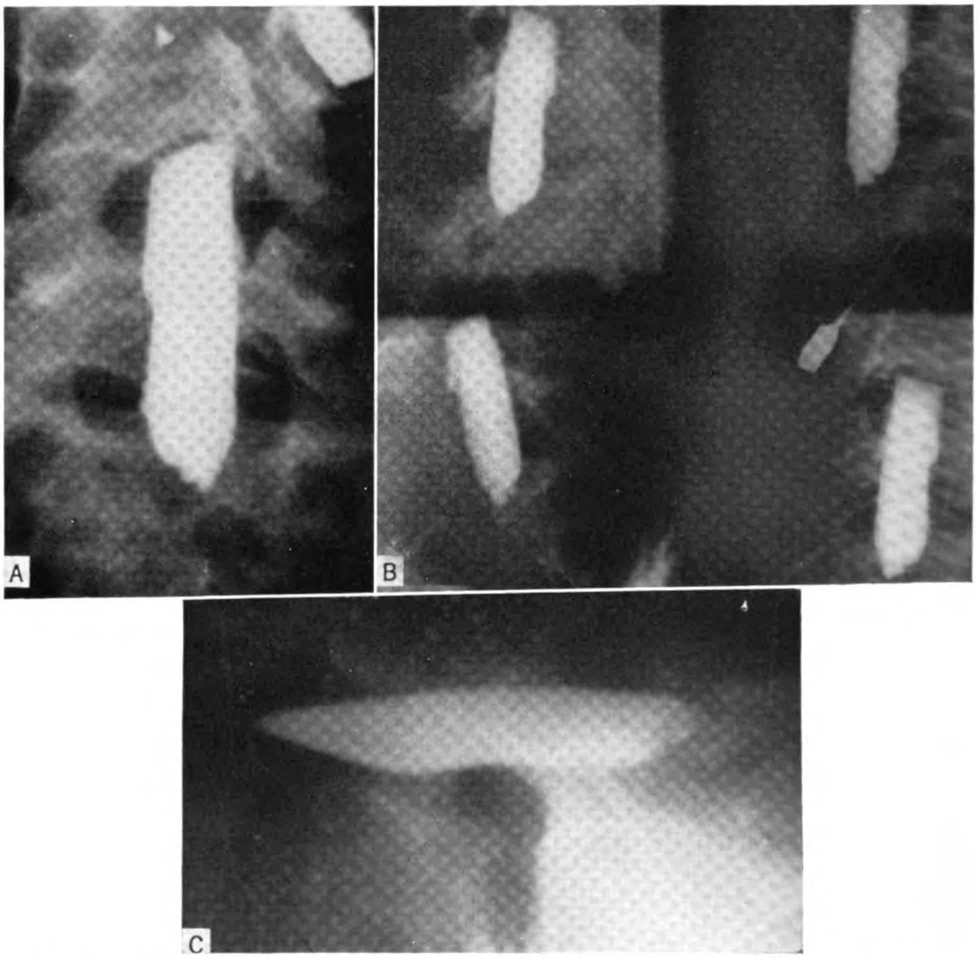

FIG. 308.—Midline disc at the lumbosacral interspace with no definite deformity of pantopaque column as seen in erect position (*A*) or in various oblique views (*B*). The transabdominal film (*C*) shows elevation of the thecal sac at the fifth lumbar interspace and proximally and distal thereto. At operation a softened disc under pressure had dissected underneath the dorsal spinal ligament, and bulged prominently when the ligament was opened.

Occasionally a horizontal, veil-like defect appears due to discal degeneration without rupture bulging against the posterior spinal ligament. Fluoroscopic study and spot roentgenograms in various oblique positions are necessary to see whether a relatively deeper indentation is present on the lateral margins of the column. Transabdominal views are required in this situation, and if these are not conclusive, right and left lateral views with the patient lying on either side should be made with the central ray passing through the midabdomen anteroposteriorly.

A fourth variety of defect, observed with massive herniation of the intervertebral disc is complete spinal canal obstruction. The entire degenerated disc with a rim of fibrocartilage may be extruded into the epidural space. It is sometimes possible to predict this when xanthochromic fluid is obtained on lumbar tap. A protein content exceeding 100 mgm. per cent is not uncommon in this situation, particularly if the extruded disc is high in the lumbar canal. With high extruded discs multiple dry lumbar taps may be encountered even though the myelographer is quite positive that the needle is in the subarachnoid space. It may become advisable to make the puncture at the second lumbar interspace and introduce only a few drops of pantopaque. Fluoroscopic examination in the erect posture will quickly disclose whether or not a block exists. Rarely it may become necessary to introduce the contrast medium through a cisternal tap.

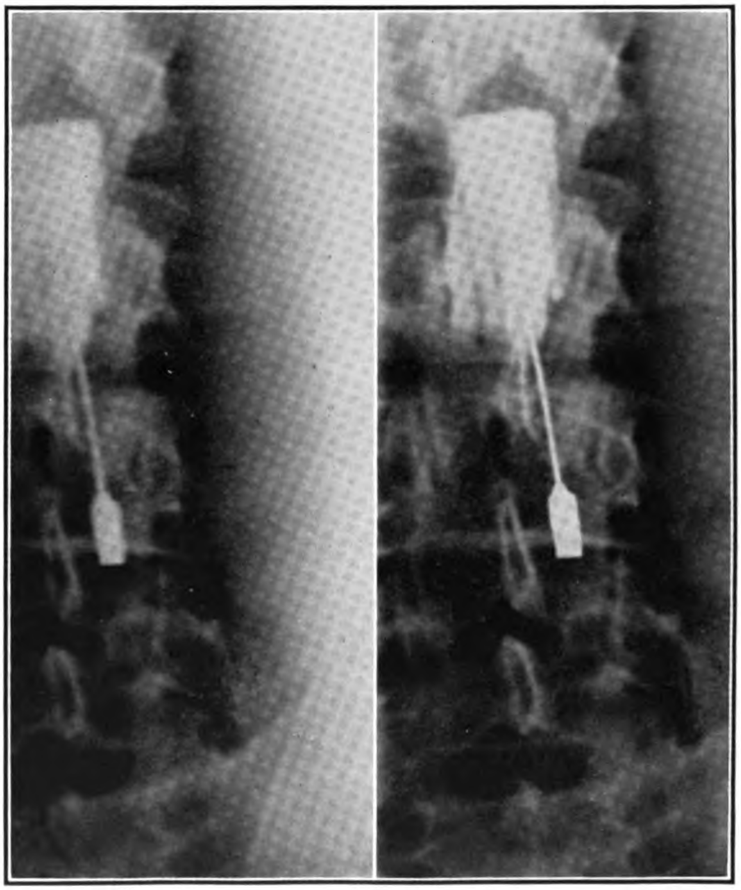

FIG. 309.—Complete block at the third lumbar interspace due to extruded herniated disc. The lumbar punctures in the third and fourth interspaces were unsatisfactory, and a good flow was obtained at the second. The linear radiolucent streaks represent prominent nerve roots. (From Epstein, B. S., Am. J. Roentgenol., **61**, 778, 1949, courtesy of Charles C Thomas, Publisher.)

An almost complete block to the flow of pantopaque may occur even when manometric measurements are normal and the cerebrospinal fluid is crystal clear. The protein content may be moderately elevated, but normal values are not uncommon. Fluoroscopic study under these conditions often discloses a lateral or posterior channel through which the pantopaque passes.

In cases with partial or complete block, and sometimes when no block at all is present, linear vertical and somewhat oblique radiolucent streaks appear within the pantopaque column both above, and sometimes below the lesion. These have been noted at operation to be due to swelling of the roots of the cauda equina. Medial displacement of nerve roots, either normal or edematous, by laterally placed herniated discs is seen relatively often. Upward displacement by midline herniated discs is best demonstrated on transabdominal films, and constitutes an important observation. It may be present in the absence of any lateral deformity.

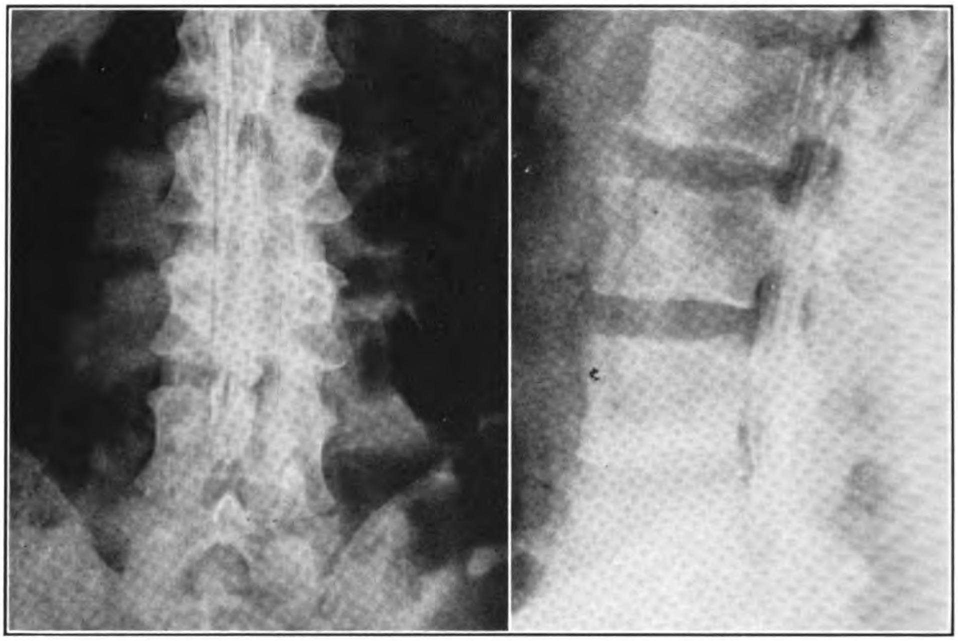

FIG. 310.—Marked swelling of the nerve roots in the cauda equina due to pressure of a large herniated disc at the fourth lumbar interspace.

By far the most frequent defects seen with herniated discs are the lateral ones, either lifting, flattening or obliterating the axillary pouch. Indentations or elongated smooth, somewhat scalloped deformities are produced by larger lesions. Complete or almost complete block is seen more often above the lumbosacral interspace than at this level.

The question may arise as to whether changes in position influence the configuration of myelographic defects. Wilson and Ilfield (1952) reported on 18 myelograms during which rotary manipulations were made to see if any change was produced in the position of the herniated disc. Thirteen had myelographic defects, and in 12 of these no change was produced. In 1 there was a slight increase in the size of the defect. Our experience is in accord with this. However, Childe (1945) mentioned that weight-bearing may considerably increase the size of a defect, and that application of traction to the feet and shoulders may reduce its size. Flexion of the spine tended to increase the size of a lesion and extension to decrease it. He also observed that in a nervous, tense patient the defect may appear larger than it did when relaxed. We have noted that a considerable variability in the appearance of myelographic defects may occur while the pantopaque flows over the lesion, so that on successive spot roentgenograms different configurations were observed. This variability in appearance was not associated with any straining or change in position, but was probably due to the extent of the envelopment of the protruded disc by the contrast medium.

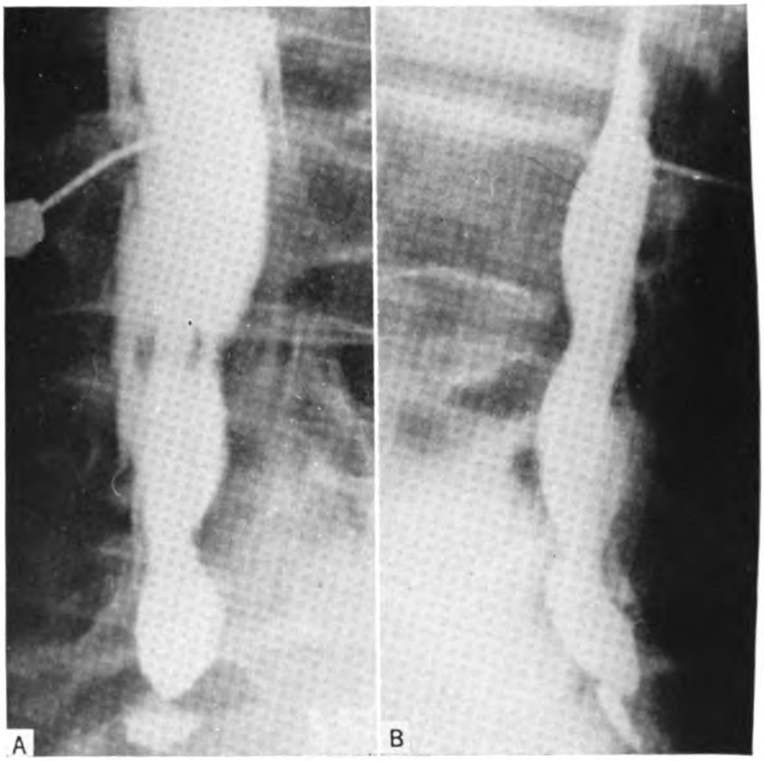

FIG. 311.—Multiple defects at the third, fourth and fifth lumbar interspaces anteriorly and on the left side as well as at the dorsal aspect of the column at these levels in a 54 year old man with progressive weakness of both lower extremities for over ten years, diagnosed clinically as multiple scloerosis. Combined multiple ridge deformities together with discal herniation at the fourth interspace and markedly thickened ligamenta flava and neural arches at operation.

The effects of postural variation on the available space in the spinal canal becomes more apparent on examinations made after filling the canal with sufficient pantopaque to opacify it up to the level of the third lumbar vertebral with the patient erect. Studies in flexion and extension indicate the effect of wrinkling of the ligamentum flavum, and the indentations so produced on the dorsal aspect of the pantopaque column occasionally are striking. However, the clinical implications of such changes must be considered with regard to the available space in the canal. If the canal is deep, and if there is no osteophytic or spondylotic ridging or bulging of softened intervertebral discs, a moderate degree of dorsal indentation is not necessarily significant. However, when the canal is less than about 1.5 cm. in its sagittal diameter, and when there is some ridging or discal bulging, such protrusions may assume clinical importance. Narrowing of the spinal canal is accompanied fairly often by thick laminæ, short heavy pedicles and some elongation and narrowing of the intervertebral foramina which can be seen on plain film examinations.

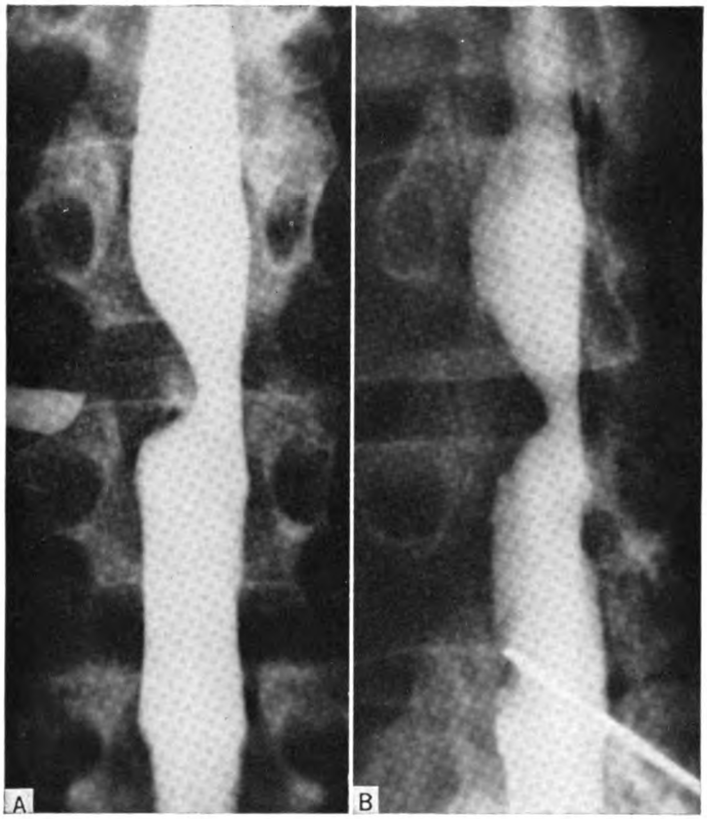

FIG. 312.—Lateral defect at the second lumbar interspace on the right side due to a herniated disc (*A*). Four months later symptoms had recurred, and a repeat myelogram (*B*) showed an almost identical defect. Recurrent herniated disc found at operation.

The concurrence of herniated discs with degenerative lesions is fairly frequent in older age groups. These bony spurs and cartilaginous ridges cannot be clearly distinguished in some cases from discal herniations. The presence of multiple ridges on lumbar myelograms is highly suggestive of degenerative changes, but the coexistence of discal herniation cannot be excluded. By the same token, a small bony protrusion may indent the pantopaque column in exactly the same way as a herniated disc. The presence of multiple ridges in the lumbar canal, particularly together with sagittal narrowing, sometimes is suspected when lumbar punctures are difficult, and often either blood-stained fluid or a poor flow of clear fluid is obtained. It is advisable under these conditions to inject only about one cc. of pantopaque and check the flow fluoroscopically before proceding. It has been my experience that removing the pantopaque from such spinal canals can be quite difficult.

Occasionally a patient who had been relieved of symptoms after removal of a herniated disc returns with complaints very similar to those present before operation. This may occur without cause or after trauma, exertion or strain. The diagnosis of a new or recurrent herniated disc must be considered, and myelographic examination becomes indicated again if further surgery is planned. Recurrence of symptoms has been observed within a few days to many years after operation.

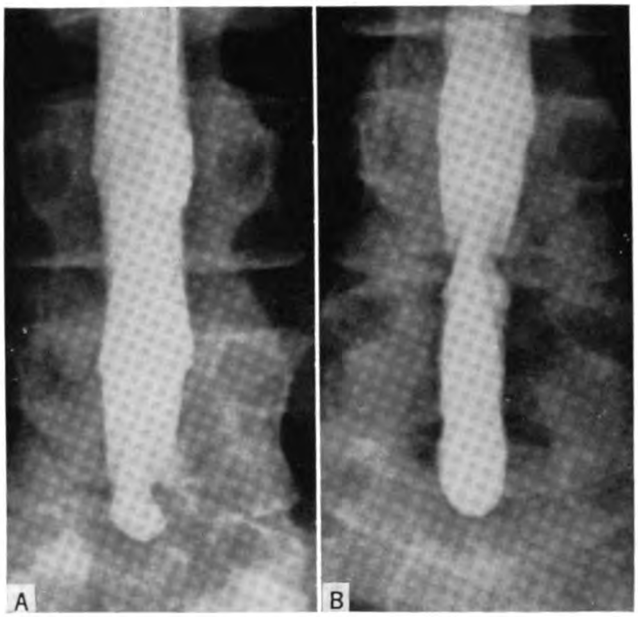

FIG. 313.—Recurrent back pain with bilateral fluctuating sciatic radiation in a 52-year-old woman who had had a herniated disc at the fourth lumbar intrespace operated upon four years ago. The present myelogram revealed a hesitation at the fourth lumbar interspace in the passage of pantopaque (*A*), and a waist-like defect when the patient was erect and the pantopaque filled the lumbar canal (*B*). Re-operation revealed a markedly thickened ligamentum flavum and a thick ridge of inflammatory tissue encircling the dura. No evidence of recurrence of discal herniation was found.

Plain film study is required to help exclude the presence of some other cause for backache and radicular pain. Among the possible reasons one must include infection or an aseptic necrosis at the operative site. Progressive narrowing of the disc space and erosive bony changes followed by sclerosis and ultimate fusion of adjoining vertebral bodies are indications of this condition.

If some pantopaque is still present in the spinal canal from a previous study, its flow should be checked fluoroscopically. Restriction of its passage and dispersal into fixed droplets may point to arachnoiditis. However, even when this is present the possibility of a recurrent or new discal herniation cannot be excluded. The addition of more pantopaque into an already inflamed area should not be undertaken unless absolutely necessary. If it is decided to do this, it is advisable not inject more than one half to one c.cm. If this does not flow freely the diagnosis of arachnoiditis may be made on the basis of the typical scattered droplets.

If the original myelographic study had been performed without leaving a useful residual, it is advisable to first inject about one c.cm. and check its flow. Only after free movement has been ascertained should the remaining quantity be added, and the examination is then continued in the usual manner. The presence of lateral or anterior defects must be regarded as evidence of recurrent or further herniations of discal material if it occurs at the site of the first lesion. Additional herniated discs sometimes occur at other interspaces, presenting characteristic myelographic malformations. The possibility of scarring incident to previous surgery also must be recognized. This sometimes deforms the dorsal aspect of the dural sac, and can be checked only if adequate quantities of pantopaque are used to permit visualization of the area immediately under the ligamenta flava. Films in hyperextension should be obtained. I have seen two cases in which scarring of the ligamentum flavum at the operative site resulted in thick bars of tissue which projected downwards towards the canal causing partial obstruction to the passage of pantopaque. Postoperative bleeding or hematoma formation likewise may be associated with arachnoidal or epidural changes causing deformity of the pantopaque column. As with primary myelography, every effort should be made to remove as much pantopaque as possible after the examination is finished.

The Necessity for and the Accuracy of Myelography.—As one reviews the literature, it becomes apparent that not all are in accord with the opinion that myelography should be a routine procedure when exploration of the spinal canal is contemplated. As early as 1941 Dandy, in a paper entitled "Concealed Ruptured Intervertebral Discs" included in the heading "A Plea for Elimination of Contrast Medium in Diagnosis." Deery (1943) believed that physical examination alone could indicate the level of a lesion and that contrast examinations were unnecessary. Hyndman (1946) regarded myelography as of little value. Scoville, Moretz and Hankins (1948) surveyed 200 operated cases in which pantopaque myelography had been performed. They stated that when myelography alone was used as a diagnostic criterion, the over-all error was in the neighborhood of 25 to 35 per cent, in contrast to the 6 to 8 per cent cited in the literature. They noted that the greatest error occurred in cases incorrectly diagnosed as normal, and mentioned that presumably normal myelograms in the face of clinical evidence of a herniated disc would be wrong three times out of four. Nevertheless, they proposed to continue to use myelography routinely on all patients suspected of harboring herniated discs, but urged conservatism in evaluating so-called normal myelograms. Lindgren (1949) also regarded routine myelography as unnecessary, and concurred with the impression that a negative myelogram did not exclude the presence of a herniated disc. Conservatism in the use of discography and myelography was urged by Lindblom (1950). O'Connell (1951) considered it unnecessary to do either a lumbar puncture or myelogram to diagnose herniated discs, since in his opinion the diagnosis could be made readily by clincal and direct radiologic study. Key (1952) advocated the use of myelography in selected cases when the clinical picture was confusing.

Hampton (1940) recorded an accuracy of 93 per cent in the myelographic diagnosis of herniated discs in 133 cases. Echline, Ivie and Fine (1945) cited instances in which the unreliability of diagnosis based on clinical signs and symptoms was corrected myelographically. They described cases in

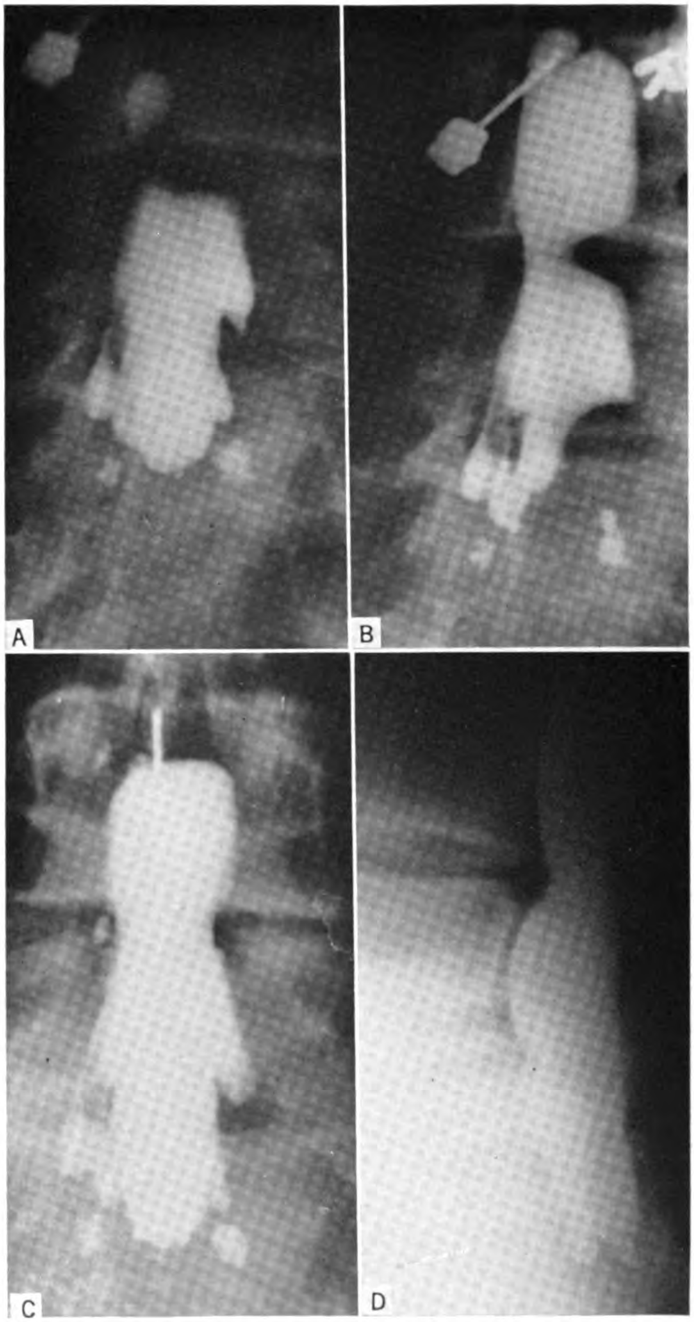

FIG. 314.—Erect myelogram with 6 cc. of pantopaque (*A*) shows a slight indentation into the left side of the column at the lumbosacral interspace. With the patient semi-erect a double defect is visible at the fourth and fifth interspaces respectively (*B*). The lesions are not as well seen with 12 cc. of pantopaque in the erect (*C*) and transabdominal projections (*D*).

which unsuspected lesions had been disclosed by this procedure. Peacher and Robertson (1945) reported their observations in 300 successive myelograms, disclosing an accuracy of 96.7 per cent in 150 operated cases. Arbuckle, Shelden and Pudenz (1945) advocated routine myelography whenever operation was proposed. It was these authors who demonstrated that a negative myelogram in a patient whose subarachnoid space measured less than 16 mm. was unreliable.

An accuracy of 89.4 per cent in the diagnosis of disc lesions by myelography was reported by Maltby and Pendergrass (1946). Grant, Austin, Friedenberg and Hansen (1948) and Colonna and Friedenberg (1949) stressed the usefulness of myelography when patients suspected of disc lesions did not respond to treatment. The dangers of overlooking spinal lesions because of reluctance to use myelography was emphasized by Svien, Dodge and Camp (1951). These authors reported an analysis of 1,245 surgical cases of herniated lumbar discs and 51 cases of spinal cord tumors which arose at or below the tenth dorsal vertebra. They found that in 20 cases of spinal cord tumor the preoperative diagnosis was herniated disc at the fourth or fifth lumbar interspace. In 7 others the possibility of a herniated disc or a spinal cord tumor was considered. The signs and symptoms in all 27 cases strongly suggested the presence of a herniated disc at the fourth or fifth lumbar interspaces, but in only 5 were the tumors located there. In the remaining 22 cases the lesions were cephalad to the fourth lumbar level. In 19 cases removal of the tumor, and delay in operation might have resulted in irreparable cord damage. Svien, Dodge and Camp cited 3 cases in which operation had been performed elsewhere, and symptoms persisted. Subsequently myelography revealed spinal cord tumors. In 2 of these spinal fusion had been done. They re-emphasized the possibility of overlooking intraspinal lesions if myelography did not precede operation.

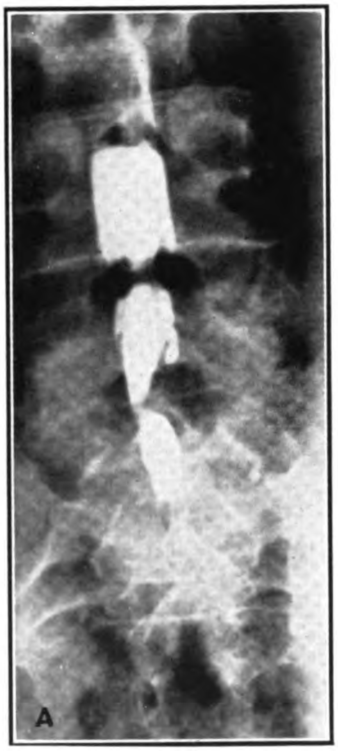

FIG. 315.—Double disc herniations at the fourth and fifth lumbar interspaces.

Leader and Rassell (1953) reported an accuracy of 86 per cent in myelographic diagnosis of herniated lumbar discs as compared with a clinical accuracy of 61.6 per cent in 500 cases. These authors found 18 cases in which myelographic defects were seen but in which no discs were protruded at operation. There were 9 with two myelographic defects in which a herniated disc was found at one level but not the other. Four cases presented myelographic defects at one level and herniated discs at another. Six had defects opposite to the lesion. Among 27 false positives surgical investigation disclosed eight bony abnormalities, of which three were seen on the roentgenograms. Five had varices and six miscellaneous lesions were found. In 8 nothing of a pathologic nature was encountered. In discussing negative explorations in 14 cases, Echols and Rehfeldt (1949) mentioned that the myelograms were normal in 10. They felt that in view of the negative explorations, it was possible that there were disc protrusions or bony spurs in the intervertebral foramina. Interestingly enough, the postoperative results of these 14 were recorded as excellent in 6, satisfactory in 2 and unsatisfactory in 6.

Both discography and water-soluble contrast myelography have proponents who favor these procedures over pantopaque myelography. After considering these technics and reviewing our own experiences with pantopaque myelography I see no reason to abandon the latter. I have frequently been impressed with the exquisite accuracy of discography, so much so that the clinical correlations become rather puzzling. It is well known that many patients with degenerative changes in the intervertebral discs are completely asymptomatic. By the same token, injection of contrast material into the discs of these people discloses passage of contrast material well outside the normal confines. Which, then, are the ones who require surgery and which should be left alone? It has been our experience that most patients operated upon for discal herniations require surgery at but one level, and are relieved of their complaints. The occurrence of multiple discal herniations and spondylotic osteophytes is not nearly as frequent, and from a clinical viewpoint, can be identified quite adequately with pantopaque studies. While it may be argued that the great majority of patients with single herniated discs might have recurrences at other levels, the fact remains that most do not. When, then, should the patient be operated upon? To us the answer is only when his clinical condition is such that there is no other way of relieving him. How much surgery should be done? My surgical friends tell me that only that which is required to relieve the offending pressure. The advisability of curetting all softened discs because they present an abnormal pattern of contrast material dispersion requires considerable thought, and at present we believe that this should not be done. Another disadvantage of discography is the fact that the examination is limited to three spaces, leaving the constant possibility that a lesion above may be overlooked. Insofar as water soluble contrast myelography is concerned, I hesitate to use it because of the necessity for spinal anesthesia, because the method limits examination to the lumbar canal, and because of possible adverse side effects (Munro, 1956).

I am convinced of the importance of myelography before exploration of the spinal canal. While it is by no means infallible, it compares most favorably with other diagnostic procedures. Considering the serious import of spinal surgery, all information should be available. In the past nineteen years, our accuracy has been between 85 and 90 per cent. I am not aware of any instances in my own experience in which myelography proved hazardous. If the procedure is skillfully done, and if the pantopaque is removed completely or almost completely, there should be no ill effects other than those which might follow spinal tap.

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Herniated Thoracic Intervertebral Discs.—Protrusions of the nucleus pulposus in the thoracic spine are uncommon. Their incidence was estimated by Love and Kiefer (1950) as occurring from 2 to 3 times per thousand cases, equally frequent in men and women. Apparently trauma plays no part in the occurrence of this lesion. The symptoms associated with thoracic herniated discs are variable, depending upon the degree of compression of the cord and nerve roots, and whether the lesion lies laterally or centrally. Symptoms vary in duration from a relatively acute onset with rapid progression requiring urgent attention to long-standing complaints which may have been present for as long as twenty years. The symptoms cannot be clearly differentiated from spinal cord tumors. In the upper dorsal area the clinical picture may simulate that seen with herniated low cervical discs. In the midthoracic spine radicular pains predominate, and in the lower thoracic area atypical symptoms referable to the abdominal viscera, the flanks and the inguinal regions appear. The usual course of spinal cord compressive symptoms including pain, tingling, burning, coldness or numbness, followed by symptoms referable to the motor tracts and sphincter control were observed in the 11 cases reported by Logue (1952). These were progressive and fairly rapid, so that the time between the appearance of symptoms and the indications for surgery was less than seven months in 8 of his patients. He observed that in 3 paraplegia developed acutely. In 2 the protrusion had apparently existed for a long time because the prolapsed disc was bony hard. It was believed that the paraplegia was due to circulatory disturbances in the spinal cord. In the other case the prolapsed nucleus was soft, and believed to be of recent origin. Symptoms referable to the legs appeared first. Additional complaints referable to one side or the other, presenting a Brown-Séquard syndrome, may be caused by lateral compresion of the cord. As a rule symptoms appeared bilaterally.

The sensory manifestations follow the pattern caused by compression of the anterior tracts, with pain and temperature sense chiefly affected. Occasionally the dermatome distribution of sensory disturbance does not correspond with the level of the lesion. Logue quoted 1 case in which the dermatome level was four segments higher than that of the spinal segment compressed. This situation also occurs in the cervical area.

Sphincter disturbances occurred in 5 of Logue's cases, and were mild. Three had slight urgency or dribbling, and in 2 others with paraplegia loss of sphincter control occurred. Love and Kiefer (1950) noted that in 5 of their 17 cases the neurologic examination was negative. They commented on the paucity of signs. In 8 of their cases the initial diagnosis was spinal cord tumor.

Investigation of the spinal fluid shows variable results, extending from completely normal observations both as to pressure and protein content to complete block with marked elevation of the protein. Normal observations are by no means uncommon. Neither of the 2 cases seen by us had manometric evidence of block or elevated protein content.

Plain film roentgenograms of the dorsal spine may reveal calcification of the nucleus pulposus, thereby affording a guide to the correct diagnosis. However, such calcifications frequently are seen in patients without symptoms, and in themselves are not acceptable as diagnostic criteria. In 5 of Logue's 11 cases the level of the calcification of the intervertebral disc corresponded with the site of protrusion. However, in another case, the level of the disc prolapse was four intervertebral spaces lower than the visible calcification. In another there were four calcified discs of which only one had protruded. In 3 no visible calcifications were present. Love and Kiefer reported that in 3 cases routine preoperative roentgenograms showed discal calcification, and in the fourth pathologic narrowing of the involved interspace was found. In 5 cases of discal herniations in the middorsal spine observed by us, the preliminary roentgenograms were normal in 2, two showed narrowing of the involved interspace, and in one a calcified nucleus pulposus was present in the involved interspace.

The location of herniated thoracic discs are fairly evenly distributed from the fourth to the twelfth interspace. Young (1946) mentioned 1 of 4 cases at the level of the third thoracic disc. Of a total of 43 cases, Logue found most to occur in the lower half of the thoracic spine. Kroll and Reiss (1951) also encountered a predominance of thoracic disc herniations in the lower thoracic spine, and observed that such lesions had a tendency to be multiple.

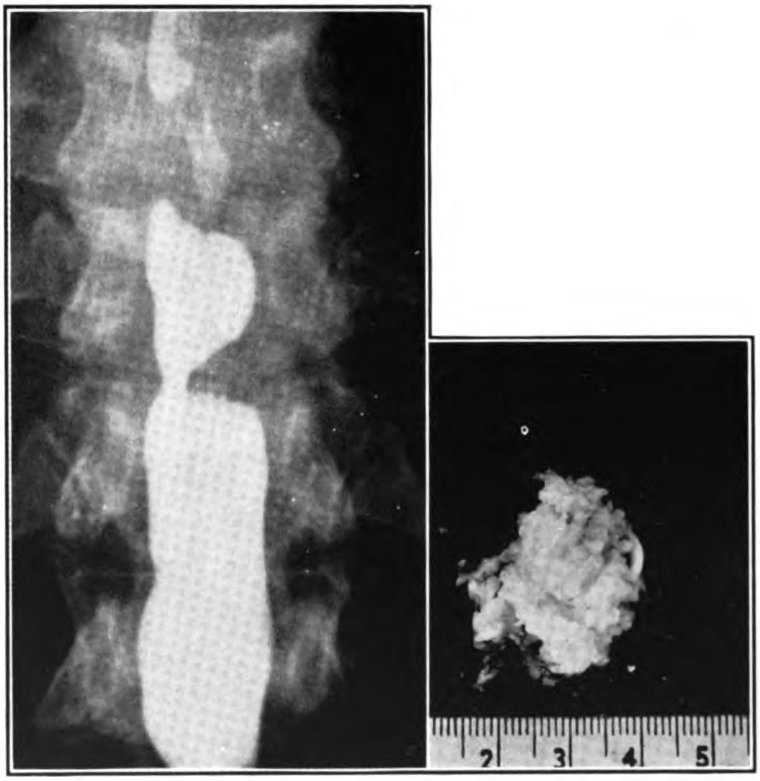

Fig. 316.—Lateral defect at the eleventh thoracic intervertebral space due to herniated disc Operative specimen. (Courtesy of Dr. Joseph A. Epstein, The Long Island Jewish Hospital, N. Y.)

Myelography is important in establishing the level and extent of the protrusion. As a rule the lumbar approach is advisable, and the most frequent change seen is a partial or complete block with a semicircular or oval defect in the head of the pantopaque column. Not infrequently some of the pantopaque passes to one side of the defect, permitting visualization of the canal above the level of the lesion. The uppermost aspect of the lesion can then be identified by returning the patient to a semi-erect position. In the lateral

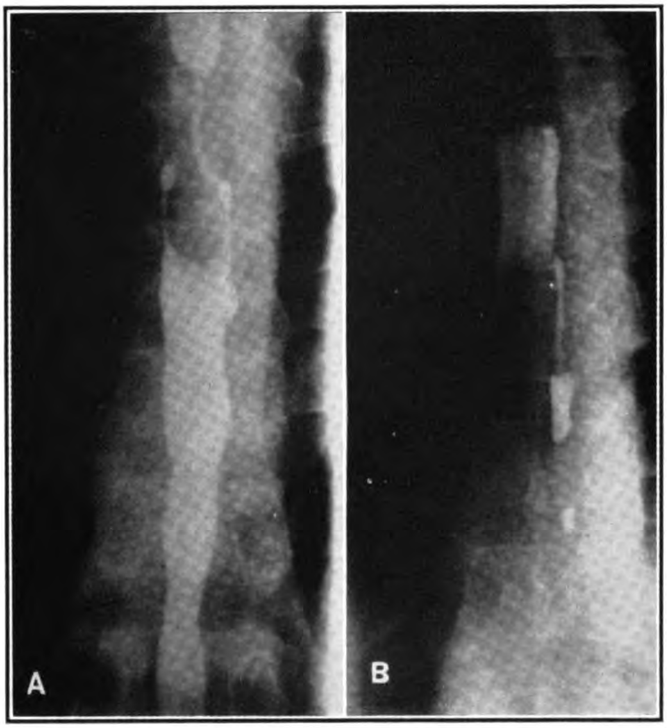

FIG. 317.—*A*, Herniated disc at the level of the sixth thoracic interspace, with pantopaque flowing alongside the disc on either side. *B*, Same case, showing the upper aspect of the defect by permitting the pantopaque to flow downwards.

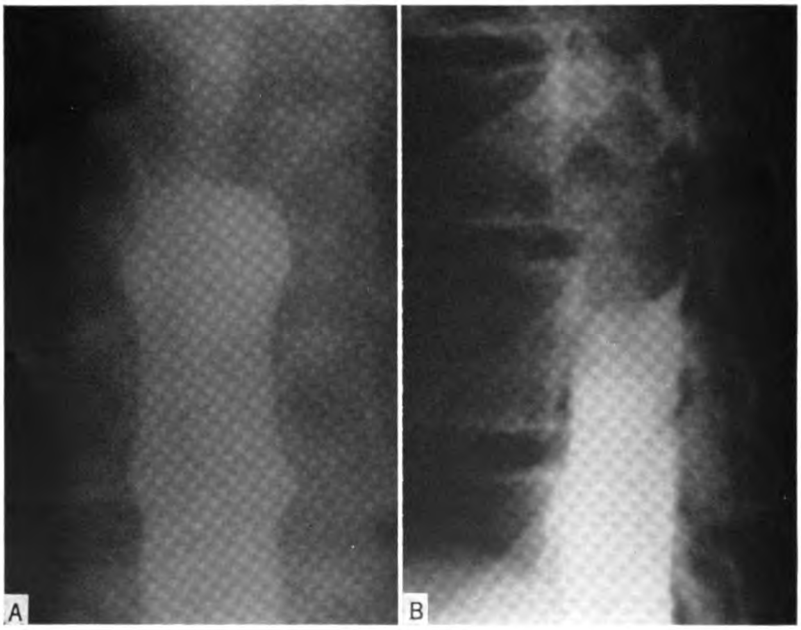

FIG. 318.—Herniated tenth thoracic disc in a 36-year-old woman with back pain for 18 months and progressive weakness. A thin trickle of pantopaque is seen at the dorsal aspect of the spinal canal (*B*). Some calcification is present in the intervertebral disc. An almost complete obstruction was noted in the frontal view (*A*).

view the pantopaque is seen enveloping the lesion, resulting in a fork-like appearance of the column at the level of the prolapse. While it is relatively easy to identify the lesion in the presence of a definite block, it is considerably more difficult to make the diagnosis when the pantopaque flows freely through the thoracic spinal canal. This was particularly well emphasized by 1 of our cases. This was a forty-three-year-old man with weakness of the lower extremities together with neurologic deficits referable to the sixth thoracic interspace. Myelographic examination showed no obstruction. However, at the suspected interspace there was a momentary hesitation in the passage of the contrast medium, with a deviation of the column to the right side resulting in a small lateral hollow. When the entire column was over the lesion it could not be identified. At operation the posterior spinal ligament was intact but bulging. When it was incised degenerated material protruded.

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Cervical Spine Herniated Discs.—This condition was first brought to attention by Stookey (1928), who reported on compression of the spinal cord due to "ventral extradural cervical chondromas." Dandy (1929) included 1 case of cervical cord compression due to a protruded intervertebral disc. In 1940 Stookey recognized that these "chondromas" were protrusions of the nucleus pulposus, and depending on the size and location of the lesion, compression was exerted on the spinal cord. Bilateral ventral pressure resulted in a clinical picture resembling a spinal cord tumor with pain, numbness, spasticity and weakness of the upper and lower extremities as well as atrophy, fibrillation and bilateral sensory disturbances. With more lateral lying lesions pressure might be exerted against half of the cord. This produces focal atrophy of the lower motor neuron type at the level of the lesion, together with spasticity and ipsilateral pyramidal tract signs of the upper motor neuron variety below the level of the lesion and dissociated changes on the opposite side. When the disc causes lateral pressure it may produce nerve root compression with radicular pain.

It was soon recognized that cervical disc herniations were less common than lumbar protrusions. However, from an anatomic viewpoint, Haley and Perry (1950) observed in 90 cadavers that cervical protrusions were twice as frequent as lumbar herniations. By 1936, Hawk had collected 16 cases of cervical herniated discs and reported an additional case. Over the years larger series have been accumulated, such as the 22 cases reported by Browder and Watson (1945), 20 cases reported by Kristoff and Odom (1947), 26 cases reported by Poole (1953) and a series of 33 verified cases out of 110 patients with cervical foraminal root compression reported by Spurling and Segerberg (1953).

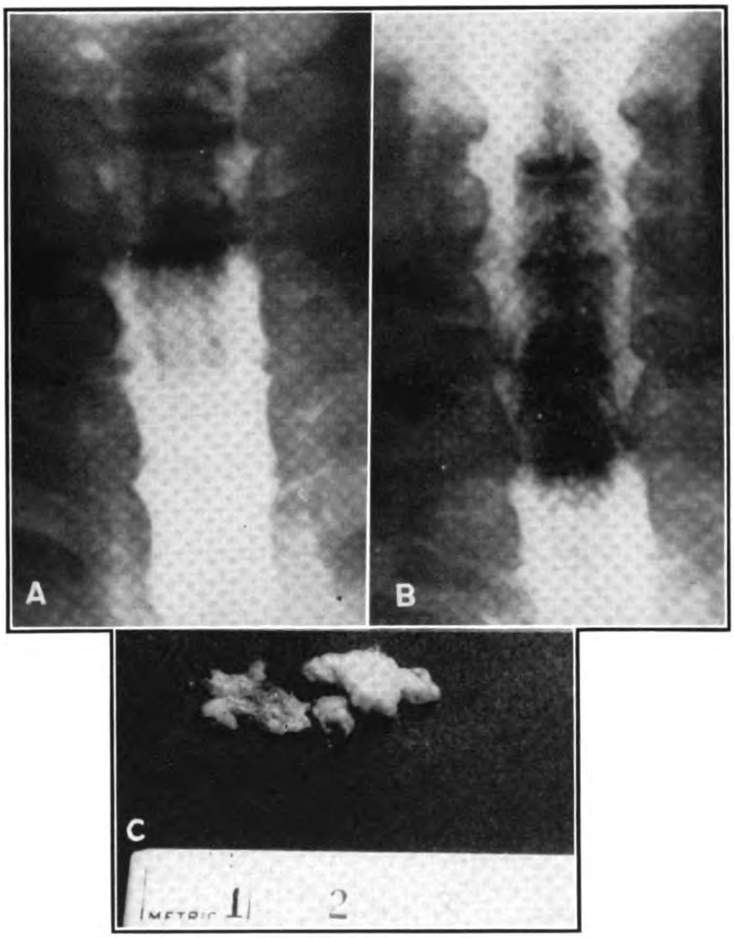

FIG. 319.—Midline herniated disc at the sixth cervical interspace in a 50-year-old man with severe left chest pain, followed by numbness, weakness and diminished reflexes in the left upper extremity after a sudden trauma. There was also a lesser degree of pain in the right arm. On fluoroscopic inspection the pantopaque met an almost transverse obstruction at the sixth interspace (*A*). When the patient was kept in the inverted position, pantopaque flowed down the lateral gutters. The spinal cord appeared to be widened (*B*). At operation a herniated disc was found in the midline and removed (*C*).

The most common site for protrusions of the nucleus pulposus of the cervical spine is between the fifth and sixth and the sixth and seventh cervical vertebræ. In the 33 cases reported by Spurling and Segerberg, 30 per cent were at the fifth cervical level and 60 per cent at the sixth cervical level. Ten per cent occurred at both the fifth and sixth cervical interspaces. These authors mentioned a personal communication from Scoville, Whitcomb and McLaurin regarding 126 verified cases of herniated cervical discs. Thirty per cent of these were at the fifth cervical level, 60 per cent at the sixth and 7 per cent at the seventh cervical levels. The remaining 3 per cent were found at the fourth cervical interspace.

In my experience there were 30 cases of herniated cervical discs. These occurred predominantly in males, with the highest incidence in the third and fourth decade. An appreciable number also were found in the decades from fifty to sixty years of age. While it was often possible to elicit a history of trauma, no specific correlation with a specific injury could be established. When a lateral lesion was present the complaint was pain in the shoulder and arm of the affected side. With central lesions and complete block of the spinal canal, as occurred in eight cases, the clinical picture was variable. Most of these patients had difficulty in moving their lower extremities together with weakness, paresthesias and pain in the upper limbs. The symptoms were referable to the dorsal funiculi, the pyramidal tracts and the lateral and ventral spinothalamic tracts. The clinical levels of neurologic disturbance were different from those anticipated from the location of the herniated discs. Root pains were relatively inconspicuous, with the entire picture suggesting a spinal cord tumor in some and degenerative disease in others. The importance of the dentate ligaments in the production of compression effects against the anterior aspect of the spinal cord was brought out by Kahn (1947). He presented stress analysis diagrams of mass lesions protruding into the spinal canal, demonstrating the role of the dentate ligaments in the distribution of pressure effects. He showed that the pressure of herniated discs on the anterior columns of the cord may produce relatively little in the way of symptoms. However, with pressure forcing the cord backwards, traction is produced against the dentate ligaments so that stress is exerted against the posterolateral columns, resulting in spasticity, weakness and disturbance of gait and balance. On the other hand, the posterior columns and the spinothalamic pathways are spared. With lesions lying to one side of the mid-line there may be compression directly against half of the cord.

The wide variation in clinical pictures has been commented upon by many observers. Raney and Raney (1948) pointed out that headache may be a prominent symptom of cervical disc herniations. Semmes and Murphey (1943) and Josey (1949) reported cases in which pain in the chest and arm simulated angina pectoris. Pain along the course of the brachial plexus, expecially in the shoulder and down the back of the arm, along the radial border of the forearm, and sometimes in the upper pectoral region was described by Elliot and Kramer (1945). They also observed paresthesia in the thumb, index and middle fingers. In some cases the patients complained of acute onset of a stiff neck. The pain was aggravated by movements of the neck and downward pressure of the head. Tenderness, weakness and wasting of the pectoralis major, triceps and extensor muscles of the wrist and fingers were also observed, together with hypalgesia over the thumb and index finger. Pain directly over the lower cervical spine is rather infrequent, although the spinous process of an involved segment may sometimes be sensitive to pressure (Michelsen and Mixter, 1944). It has long been recognized that herniations of the nucleus pulposus, particularly large ones in the midline, may produce syndromes indistinguishable from spinal cord tumors or obscure neurologic disturbances such as syringomyelia, multiple sclerosis, amyotrophic lateral sclerosis or combined system disease (Epstein and Davidoff, 1944; Bucy, Heimburger and Oberhill, 1948; Strully, Gross, Schwartzman and von Storch, 1951). Rarely involvement of the descending trigeminal tract may be seen with herniation of the fourth cervical disc (Elvidge and Li, 1950).

The cerebrospinal fluid changes associated with herniations of the cervical discs are variable. Manometric observations may be entirely normal except in those cases in which significant block exists. It is not uncommon to obtain normal manometric readings in the presence of an almost com-

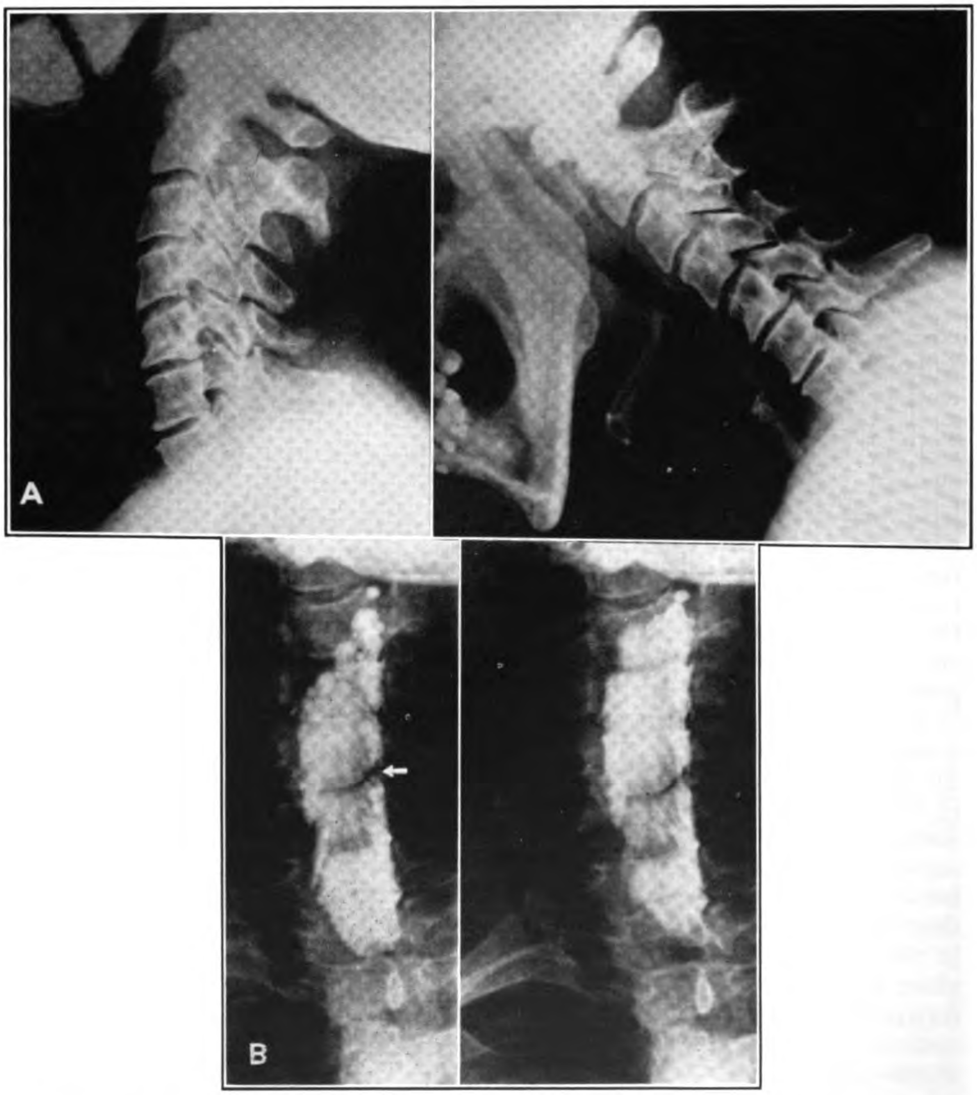

FIG. 320.—*A*, Lateral flexion and extension cervical roentgenograms showing limitation of motion. Spondylosis is present at the fifth and sixth cervical vertebræ. *B*, Same case, cervical myelograms. A small lateral defect (arrow) is present on the left side at the fifth interspace. A lateral nuclear extrusion with adjacent bony sclerosis was found at operation.

plete cervical spinal canal block as demonstrated on myelograms. The cerebrospinal fluid protein content may vary from normal values to considerably elevated figures when significant compression exists.

The cerebrospinal fluid circulation may be impaired because of intrusions into the spinal canal from spondylosis or from herniated discs. Morton (1936) demonstrated 3 cases in which manometric measurements showed subarachnoid block when the patients' heads were extended to the point where they complained of numbness in the fingers. We, too, have observed transitory pantopaque block while the head was so maneuvered during myelography in patients with large herniated discs or prominent ridges. Taylor (1953) presented myelograms demonstrating interruption of the pantopaque column on hyperextension of the neck because of wrinkling of the ligamentum flavum.

Because of anatomic conditions, protrusion of the nucleus pulposus of the cervical spine is usually somewhat different than seen in the thoracic or lumbar regions. Of considerable importance is the fact that the cervical spinal cord almost completely fills the available space in the canal. The nerve roots emerge horizontally, entering the intervertebral foramina almost directly. The amount of epidural fat is scant. The cervical nerve roots are relatively large, and the intervertebral foramina are small, so that direct compression against the bony walls may result from relatively small protrusions. The posterior spinal longitudinal ligament expands opposite each intervertebral disc, but does not reach the lateral margins. The annulus fibrosus is weakest at the posterolateral margins, so that rupture at these points is facilitated. The dentate ligaments on both sides fix the spinal cord, and their importance in the resultant effects of pressure anteriorly has been mentioned before. The intervertebral discs in the lower cervical spine are small. Bull (1948) observed that the volume of the cervical disc nucleus pulposus varied from 1.0 to 1.4 c.cm., as compared with lumbar discal contents of up to 10 c.cm.

The mobility of the cervical spine is a significant factor in damage to the intervertebral discs. Herniations may occur acutely as a result of hyperflexion injuries, and may be associated with compression fractures of the vertebræ or minor derangements of the small joints. The nucleus pulposus may protrude through a weakening of the posterior spinal ligament, so that the nodule projects into the anterolateral aspects of the spinal canal, compressing the cord laterally, or the nerve roots. This change also takes place as part of a degenerative process with no gross changes on plain film roentgenograms. More frequent is the appearance of narrowed intervertebral discs and spur formations at the uncovertebral joints directed anterolaterally and towards the intervertebral foramina. It often is very difficult to ascertain whether the primary condition is a degenerative spondylosis or a protruded nucleus pulposus. In any event, either condition is primarily a discogenic process, probably representing different phases of the same physiopathologic process, the discogenic disease being the primary lesion of deforming spondylosis, the bony proliferation being an attempt to heal the pathologic disc by physiologic immobilization.

The clinical and radiologic aspects of spondylosis with bulging and spurring of the cervical intervertebral discs were considered in Chapter 4. The compressive effects of these ridges are more prominent in the cervical than in the lumbar spine. Ridging of the thoracic intervertebral discs is associated with intraspinal pressure effects infrequently. Narrowing of the cervical spinal canal reduces the available space, so that minor ridges produce disproportionate neurologic effects. Thickening of the neural arches, particularly of their posteroinferior margins, together with hypertrophy of the ligamenta flava, likewise produce pressures which may be of great importance in narrow canals. Changes in the neural arches and ligamenta flava affect the dorsal aspect of the cervical cord, and pressures are augmented when the ligamenta flava are wrinkled inwards on extension of the neck. If spondylotic ridges are also present, they may act as anvils against which the anterior aspect of the cord is pressed. The movement of the spinal cord and nerve roots during activity enhances the pressure effects of these enveloping structures.

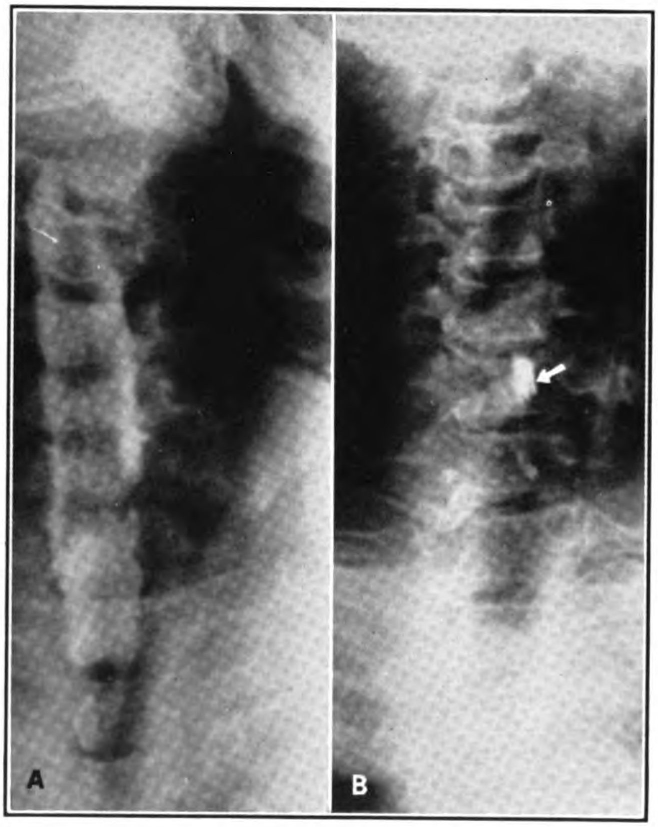

FIG. 321.—*A*, Cervical myelogram with a small lateral defect at the sixth cervical interspace on the left side due to herniated disc. *B*, When the patient was returned to the erect position, a small fleck of pantopaque is caught just above the herniated disc (arrow).

Differentiation between a herniated cervical disc or osteophytic changes affecting a single interspace sometimes is difficult even with myelographic evidence. The presence of multiple ridges points more towards spondylotic change. The importance of correlating clinical and radiographic changes is again emphasized because of the frequency with which apparently extensive bony changes are unaccompanied by symptoms, especially in the middle and later age groups. Herniated discs or single spurs occur at levels which apparently are normal on plain film investigations. In these individuals myelography is required for accurate diagnosis, but should not be utilized unless surgery is under consideration.

The classical radiologic picture associated with cervical discal herniations included straightening of the usual cervical lordotic curve, limitation of flexion and extension, spurring and discal narrowing. It is not at all infrequent to find all of these without symptoms, and on the contrary, to encounter patients with severe symptoms in whom the plain film examinations are relatively normal. Cervical discography has been advocated by Cloward (1959). I have had no experience with this method, but it appears that the same objections raised in consideration of lumbar discography apply to the cervical adaptation.

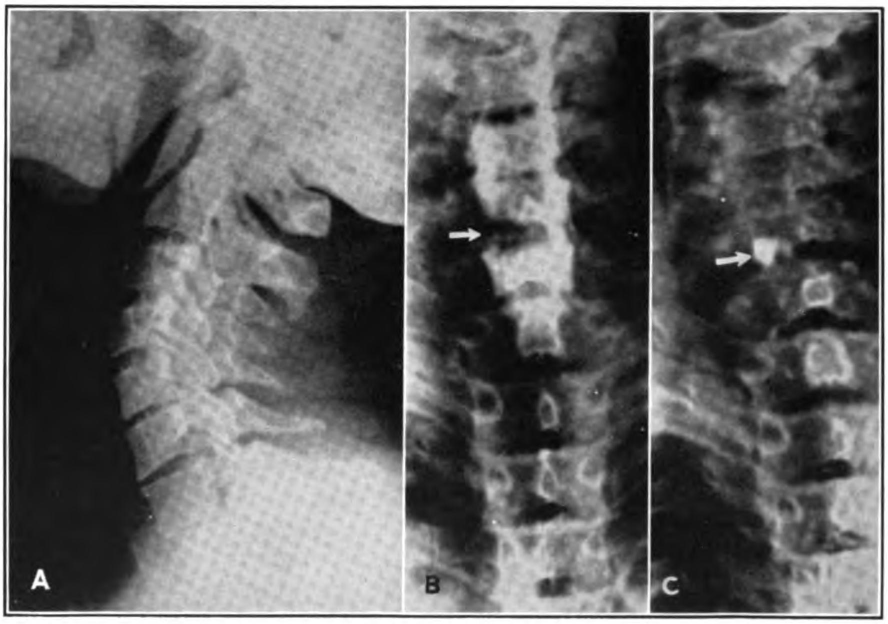

FIG. 322.—*A*, Lateral cervical roentgenogram. There is narrowing at the fifth and sixth interspaces. *B*, Same case. A rather large defect is present on the right side of the pantopaque column at the sixth interspace (arrow), due to a herniated disc. *C*, Same case. When the patient is returned to the erect position a fleck of pantopaque remains just above the herniated disc (arrow).

Myelography is of prime diagnostic importance in the identification of cervical disc protrusions, but should be used only when symptoms warrant the consideration of surgery. With proper care, myelography can be performed with little danger of pantopaque entering the cranial cavity. A proper cervical myelogram should visualize the entire cervical canal up to the foramen magnum to avoid missing lesions in the upper portion of the spinal canal.

Depending upon the location and extent of the protrusion, myelography may reveal only a small lateral defect obliterating the axillary pouch, together with a small oval or semicircular defect in the pantopaque column. With larger lesions, the defect becomes more apparent, and may produce partial or complete block, resulting in a U or L shaped deformity in the head of the pantopaque column. Multiple defects should be sought for possible multiple lesions. The effect of extension and flexion of the neck on the pantopaque column should be gently and carefully determined.

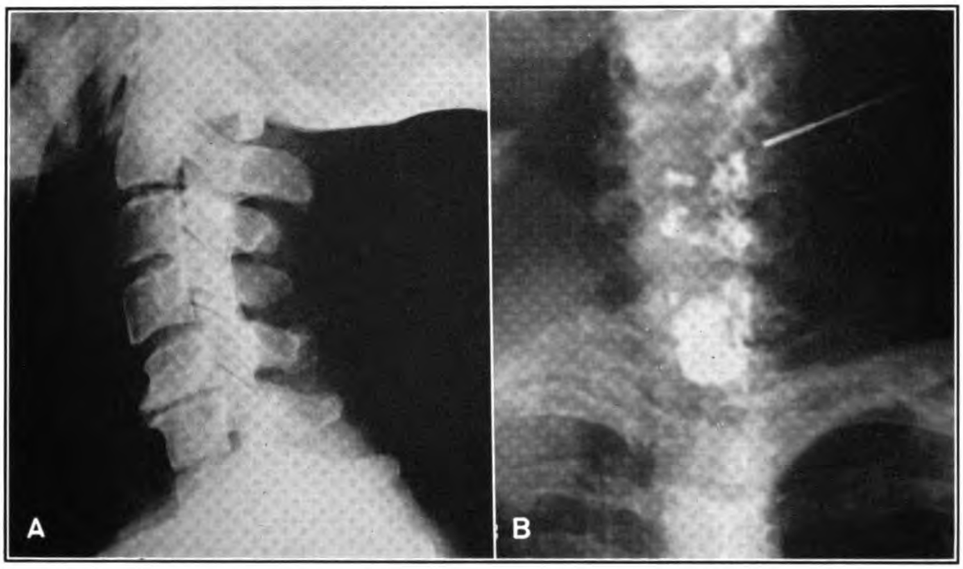

FIG. 323.—*A*, Lateral cervical roentgenogram showing straightened cervical curve and narrowing of the fifth cervical interspace with spondylosis of the fifth and sixth cervical vertebræ. *B*, Same case, cervical myelogram. A large defect causing partial obstruction is seen at the sixth cervical interspace, one interspace lower than the spondylitic changes seen on the plain film.

In hypertrophic spondylosis a corrugated appearance of the pantopaque column appears, which might be termed a "wash-board" pattern. Here the column is broken up into ladder-like ridges because of the indentations of the dorsal cartilaginous protrusions from the intervertebral discs. These sometimes are bony in consistency, and frequently are a combination of both cartilage and bone. The number of these ridges may vary from 1 to 6, and cause considerable difficulty in evaluating symptoms. In some cases it was found necessary to remove the neural arches of practically the entire cervical spine.

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ANTERIOR DISCAL PROTRUSIONS

In discussing herniations of the intervertebral discs, attention is usually directed towards symptom-producing posterior protrusions. There is an equally large, and perhaps even a larger group of discal malpositions associated with discogenic disease in which displacement of nuclear materail occurs anteriorly, superiorly or inferiorly.

Batts (1939) noted 3 instances of anterior protrusion of the nucleus pulposus in 50 cadaver specimens with herniated discs. His Figure I depicted a thinned disc with the nucleus pulposus extruded anteriorly, where it was covered with osteophytic proliferations and bony bridging between the anterior portions of the vertebral bodies. The bony changes were considered secondary to the discal alterations. Schmorl and Junghanns also demonstrated on anatomic preparations that the so-called "persistent epiphyses" or "intercalary bones" seen on the anterosuperior aspects of the vertebral bodies represented another form of discal extrusion separating a small fragment of bone from the body of the involved vertebra. Association of such changes with back pain in eight cases was reported by Hellstadius (1947).

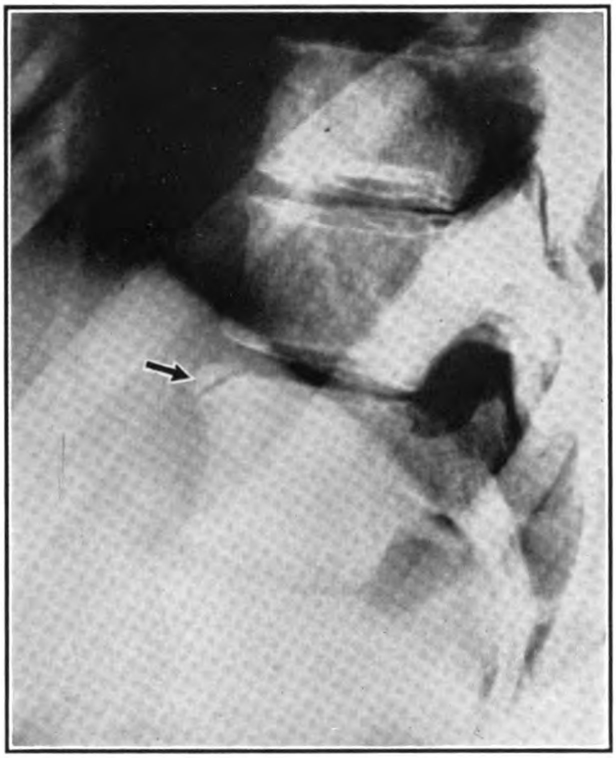

FIG. 324.—Small, triangular bony fragment at anterosuperior aspect of the first lumbar vertebra (arrow), miscalled persistent epiphysis or intercalary bone. This has been demonstrated as being due to anterior discogenic disease with forward nuclear protrusion.

By means of discography, Lindblom (1951) demonstrated on 3 anatomic preparations and 5 living patients that opaque medium entered the fissure lines separating the intervertebral discs from the marginal bony fragments, confirming the work of Schmorl and Junghanns *in vivo*. Anterior herniations of the nucleus pulposus revealed by discography was also reported by Cloward and Buzaid (1952), who commented that discograms might reveal rupture of intervertebral discs in single or multiple directions, anteriorly, posteriorly, vertically and posterolaterally. Cloward (1952) also reported a case in a forty-two-year-old man with chronic low-back pain in which discograms showed anterior herniation of a large disc fragment.

Lindblom (1951) reported that extension of diodrast during discography into an anteriorly ruptured first lumbar disc had caused severe abdominal pain. A similar reaction took place in 2 other patients without bony separation from the vertebral body. He mentioned that Grill (1951) had directed attention to the possibility of abdominal pain arising from anterior discal herniations.

Leavens and Bradford (1954) reported a case in which, after removal of a herniated nucleus pulposus, their rongeurs had passed into the interior of the disc, and then with minimal pressure went through a fissure in the anterior aspect of the annulus fibrosus. They believed this might explain abdominal vascular accidents, and cited 2 cases from the literature in addition to 12 of which they had personal knowledge in which abdominal aortic injuries terminated fatally because of hemorrhage. Smith, Hughes, Sapp, Joy and Mittingly (1957) reported two patients with high output circulatory failure due to arteriovenous fistulas following surgery for herniated discs. They noted that nine cases had been reported, five of which had been recognized only after the fistulas had resulted in circulatory failure. The injury to the vessels occurs during the removal of discal cartilage, when injudicious manipulation may result in injury of vessels anterior to the disc. I recall one case recently in which the patient expired on the operating table because the abdominal aorta was torn by the pituitary rongeurs during the procedure. A long time may elapse between the creation of an arteriovenous fistula and its radiologic or clinical diagnosis. Radiologically the lesion may be identified by means of aortography.

A rare example of anterior thoracic discal protrusion causing slight dysphagia and a defect of the esophagus because of extrinsic pressure was reported by LaSorte and Brown (1959).

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Complications of Myelography

As stated before, myelography in competent hands is a safe and reliable diagnostic procedure. Nevertheless, untoward effects have been reported and are worth reviewing. There were no fatal incidents in my experience which might have been attributed directly or indirectly to myelography. In approximately 15 per cent of cases, including those done as ambulatory patients, headaches were a source of annoyance. The onset of headache varied from a few hours to as long as ten days after myelography had been performed. These were often relieved by keeping the patient recumbent. It did not seem to make any difference whether the patient was absolutely quiet after myelography or not, nor was there any beneficial effects from various medications, forced fluid intake, caffein or ergotamine tartrate. In some the pain was aggravated by movement of the neck or sitting up suddenly, and in a few nuchal rigidity and a picture of meningismus appeared.

Another unpleasant concomitant of myelography was the persistence of low-back pain, occasionally accompanied by sciatica. These ill effects result from the lumbar puncture itself rather than from the pantopaque. This is borne out by the great similarity to complaints which follow diagnostic lumbar tap without the injection of contrast medium. Childe (1945) believed that even a drop of water on the syringes or needle might cause untoward meningeal reactions. Shipp (1953), reporting on 1800 cases of pantopaque myelography, mentioned that no significant complications attributable to the contrast substance had been encountered. In some of his cases the bulk of the oil was fixed by adhesions in as short as twenty days after myelography. He was of the opinion that such adhesions probably were the effect of actual infection introduced at the time of puncture rather than from the agent itself. Reactions in the form of meningismus and elevation of the cerebrospinal fluid cell count, and sometimes total protein elevations, occurred in 3 of the 100 cases reported by Schnitker and Booth (1945). In 10 cases with retained pantopaque in amounts varying from $\frac{1}{10}$ to 2 c.cm., lumbar puncture done at intervals of from one to four days later showed that reactions, when present, were not directly proportional to the amount of retained material. Seven of the 10 had no spinal fluid changes. In the remaining 3 there was headache with meningismus, generalized aches and fever which subsided in seventy-two hours.

Reactions attributed to pantopaque have been described. Tarlov (1945) reported a thirty-seven-year-old male who complained of severe headache, stiff neck and fever of 100.8° F. five hours after the instillation of 3 c.cm. of pantopaque. The next day his temperature rose to 102° F. One day later, when an interlaminar exploration was done, considerable whitish, soft stringy exudate was seen adherent to the arachnoid and the cauda equina. Histopathologic examination of the exudate disclosed a meshwork of fibrin strands with imbedded leukocytes, lymphocytes and plasma cells. No organisms were cultured.

Bering (1950) re-examined 25 of 106 patients who had had up to 6 c.cm. of pantopaque injected into the subarachnoid space without removal of the medium after myelography. These examinations were performed at intervals of from six months to between two and three years. None of these patients had signs or symptoms which might be attributed to the pantopaque. In approximately half these cases the pantopaque droplets had become fixed in the subarachnoid space, indicating a reaction at these points. Fixation of a large quantity of pantopaque was observed in 1 case only, while in the others the residual was freely movable. We have seen patients who had pantopaque injected as long as nine years before re-examination, and free movement of pantopaque within the spinal canal was the rule rather than the exception. However, when the pantopaque had entered the cranial cavity the droplets remained fixed in the basilar cisterns and over the brain convexities. We could not establish any clinical syndrome with this situation. One patient who had had a myelogram about ten years before returned with pain in the neck extending up the base of his skull. This was attributed by competent observers to the pantopaque in his head. However, further investigation disclosed a perineurial fibroblastoma at the second intervertebral foramen. His complaints were relieved after operation.

The impression that pantopaque is entirely innocuous, however, must not be assumed from the above statements. There is experimental evidence that in certain instances inflammatory reactions may follow the retention of pantopaque in the subarachnoid space. Schurr, McLaurin and Ingraham (1953) described an arachnoidal inflammatory reaction after injection of pantopaque into the subarachnoid spaces of dogs. Even though the amounts used were relatively greater than employed in myelography, the inflammatory changes indicated the necessity of preventing pantopaque from entering the cranium. Erickson and van Baaren (1953) reported a thirty-three-year-old man who expired fifteen months after myelography because of an exudative adhesive arachnoiditis producing obstruction to the fourth ventricle and the basilar cisterns. Necropsy revealed that the basal surfaces of the pons and medulla were covered with greenish-grey exudate. Tortuous dilated blood vessels and a fibrinous exudate were spread over the lower thoracic portion of the cord. That this reaction was due to pantopaque was indicated radiologically, by chemical determinations of iodine in the exudate and by microscopic studies.

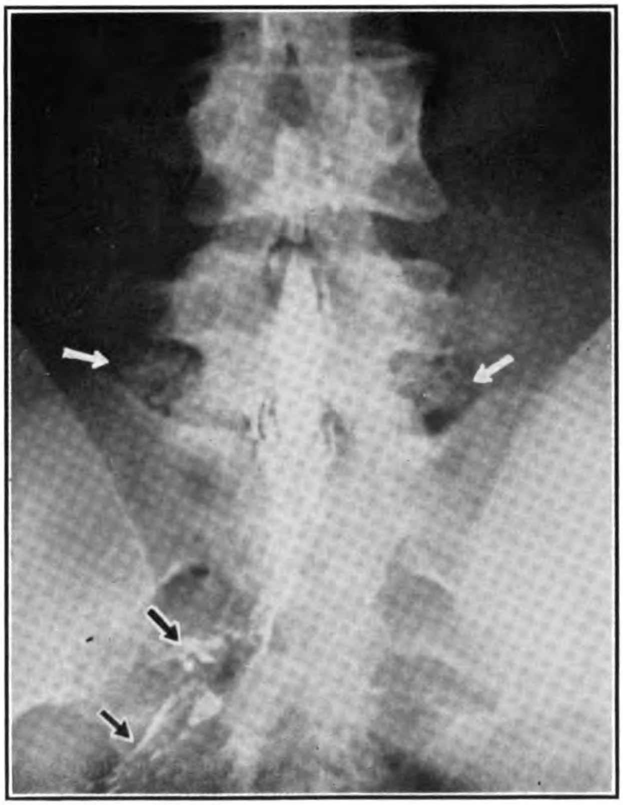

FIG. 325.—Pantopaque entering the venous tree lateral and below the main column (arrows). Within half a minute or less the entire column vanished.

Instances of intravasation of pantopaque into the spinal veins during myelography are infrequent. These may be quite dramatic, as exemplified in 1 of our cases in which the pantopaque disappeared from view during fluoroscopy. For one moment there was a network of fine and fairly large vessels filled with pantopaque, which then vanished. I was fortunate enough to obtain a spot film of this occurrence (Fig. 325) of which the patient was completely unaware. Such cases were reported by Hinkel (1945), who described a star-burst extension of pantopaque in all directions when his patient coughed. Within fifteen seconds no oil remained in the subarachnoid space, and a broad column formed to the right of the lumbar spine. His patient complained of some tightness in his chest, but no other untoward effects were noted. Another myelogram on the same patient seventeen days later was normal. Fullenlove (1949) observed similar changes, and noted that the opacification of the venous system disappeared within thirty seconds. Steinbach and Hill (1951) reported a case of venous intravasation in a twenty-eight-year-old woman in whom 9 c.cm. of pantopaque had been used. A bloody return was first obtained on trying to remove the pantopaque. A second lumbar tap was done and 1.5 c.cm. was aspirated together with bloody fluid. The patient then complained of slight chest pain and a nonproductive cough. The next day she suddenly felt sick, perspired profusely and vomited repeatedly. A chest film showed multiple small fine reticular densities in the lung parenchyma. These had disappeared by the time the patient was seen again twenty-three months later. These authors estimated that from 4 to 7.5 c.cm. of pantopaque must enter the venous circulation to show up in the lungs.

Extradural extension of pantopaque occurs every now and then (French and Strain, 1947), and is easily identified by the opaque medium along the course of the several nerves and about the spinal canal. Instead of a smooth, homogeneous smoothly flowing column one encounters an irregular, streaky, poorly flowing distribution. Within a few hours, and sometimes within a few minutes, pantopaque extends along the nerve roots to, and often beyond the intervertebral foramina. Epidural extension occurs through a small rent in the dura, and is often prominent in the lower lumbar area. The pantopaque may then extend as high as the cervical region. Repeated examinations show no appreciable absorption of pantopaque, nor are there to my knowledge untoward effects from such extensions. Occasionally extradural extension may be seen on films made several days after myelography if some pantopaque has been left in the spinal canal. This, too, occurs because of passage through a small dural tear.

Subdural instillation of pantopaque may be recognized by the unusually slow movement of an apparently smooth column. In such instances the pantopaque is situated in the space between the dura and the arachnoid, and dissects its way between these layers by gravity as the patient is placed in various positions. Sometimes the subdural space, which normally is only a potential space, may be increased greatly in depth after the entrance of cerebrospinal fluid following a previous lumbar puncture. If myelography is done soon thereafter, the pantopaque may be injected subdurally rather than into the subarachnoid space. Removal of the pantopaque under such circumstances is most difficult. I have observed this in a few cases, and no untoward effects followed, although it was impossible to go on with the examination. One might suppose that if a lesion were present, it might have deformed the subdural pantopaque column.

Luce, Leith and Burrage (1951) reported 2 cases with evidences of noninfectious meningitis after the use of pantopaque after intradermal skin testing with the same material. Both patients had a history of atopic ragweed hay fever confirmed by positive tests. The delayed appearance of the meningitis was attributed to the previous skin testing as an allergic phenomenon.

Another rare complication was described by Wright (1949), who observed paralysis of the external rectus muscles of the right eye five days after myelography, with spontaneous recovery.

At this point the advantages of removing pantopaque as completely as possible after myelography hardly need recounting, even though the possibilities of reactions are remote. If nothing more is accomplished than to guard against possible abuses of a situation, the requisite skill in removing pantopaque after myelography should be acquired by all who perform this examination. However, even in the most skillful hands, conditions will arise in which it may become almost impossible to remove the pantopaque.

Untoward effects of the mechanics of lumbar puncture and removal of pantopaque have been attributed to myelographic examinations. Among these are included the introduction of infection and the irritative effects of pushing a spinal needle through the dura. Another cause which is still insufficiently recognized is the effect of vigorous aspiration of pantopaque. If the needle tip sucks up a nerve root a considerable amount of pain may follow repeated efforts at aspiration. Recently I saw a 35-year-old patient who was complaining of pain on the side opposite that which had brought her for examination. A review of her myelograms showed that the physician who had done the examination had introduced the needle at the second lumbar interspace laterally. In all probability her pain was due to vigorous aspiration for removal of the pantopaque which, incidentally, was unsuccessful. The pain had persisted for more than two weeks after the examination, and followed a classical radicular pattern. It is our practice to withdraw the lumbar puncture needle if there is difficulty in obtaining a good return of pantopaque, and to reinsert it at the lumbosacral interspace.

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Chapter

8

Diseases of the Spinal Cord and its Coverings

INFLAMMATORY DISEASES

ARACHNOIDITIS

Arachnoiditis presents a very variable clinical picture. The condition may be inaugurated by pains referable to nerve patterns. When lumbar areas are affected, the history of back pain and sciatica may be intermittent, and suggest the diagnosis of a herniated disc. When the disease occurs in the cervical or thoracic spinal canal, its onset may be relatively acute and severe, and simulate a tumor within the spinal canal.

Many causes have been attributed to arachnoiditis. Among these are included the various inflammatory diseases such as meningitis (Mackay, 1939), pneumonia, typhoid fever, and virus diseases (Stookey, 1927). Under such conditions the disease may be of a low grade intensity and associated with long periods of minor symptoms with exacerbations and remissions. Arachnoiditis may be associated with trauma, probably incident to bleeding within the spinal canal. In this connection the experiments of Bagley (1928) are of interest. He injected the blood of dogs into the subarachnoid space by way of the cisterna magna, and produced frequent meningeal reactions in response to a few cubic centimeters of blood. In most cases this reached the state of proliferative meningitis, and he was able to cite clinical cases of a similar nature. Nelson (1943) reported the case of a fifty-year-old man with thrombopenic purpura with arachnoiditis, and postulated that meningeal bleeding occurring during convulsive episodes may have preceded sterile meningitis, which went on to fibrotic organization and cavitations. At necropsy, the inflammatory process began at the midthoracic region and extended down to the cauda equina. There was marked thickening of the dura with adherence of the nerve roots of the cauda equina. The cord above the lesion was normal, but below sections revealed numerous cavities in the dorsal and lumbar regions. The normal architecture of the cord appeared to be markedly distorted. In one of our patients with a bleeding aneurysm of the circle of Willis arachnoiditis appeared about six months after an acute episode, at which time bloody fluid had been obtained from the lumbar spinal canal. Hypertrophic interstitial polyneuritis due to hyperplasia of the sheath of Schwann may be identified radiologically by myelographic demonstration of block together with thickened nerve roots (Lewtas and Dimant, 1957). Circumscribed local arachnoiditis produces cystic lesions which behave like tumors. These will be considered under the heading of arachnoid cysts.

Arachnoiditis with calification is infrequent. It has been observed together with a vascular spinal anomaly and subarachnoid hemorrhage (Gatzke, Dodge and Dockerty, 1957). Calcifications producing multiple myelographic defects in the lumbar area were seen on myelograms by Faeth

(1958). At operation these stones were adherent to the roots and their tiny vessels, and apparently did not arise from the arachnoid. No cause could be established for these stones. However, it is not infrequent to find thin calcific plaques a few millimeters in diameter in normal meninges at post-mortem in elderly patients.

Arachnoiditis has also been associated with continuous caudal anesthesia. Peacher and Robinson (1944) described 2 cases, 1 of which was associated with a secondary osteomyelitis, while the other had either an epidural or subdural abscess which responded to chemotherapy. Kennedy, Effron and Perry (1950) reported on the occurrence of inflammatory lesions following spinal anesthesia. Rosenbaum, Long, Henchey and Trufant (1952) described several cases of paralysis following saddle block anesthesia in obstetrical patients. Progressive arachnoiditis appeared in a patient seen by us who had had spinal anesthesia for a subtotal gastrectomy ten years before he came for help. The onset of symptoms had been gradual over a period of about five years.

The irritative effects of lipiodol on the meninges with the production of a mild form of arachnoiditis was demonstrated experimentally in dogs by Craig (1942), and was discussed from the clinical viewpoint in the chapter on myelography. Mention also has been made of reports of arachnoiditis following the use of pantopaque in the discussion of complications of myelography. Walker (1947) described 2 cases of arachnoiditis after the intrathecal administration of penicillin. He stated that such reactions usually occurred only with large amounts of penicillin, but that when smaller doses were used this complication was rare. In his second case, cisternal and lumbar myelography showed changes characteristic of arachnoiditis, and this was confirmed at operation.

With the advent of frequent operations for herniated discs it was noted that occasionally these lesions were accompanied by inflammatory changes in the leptomeninges. French (1946) reviewed 200 such cases, and found 13 with arachnoiditis, 8 of which had concomitant herniated discs. The remaining 5 were early cases, and protrusions may have been overlooked. The association of arachnoiditis with spinal cord surgery was mentioned by Smolik and Nash (1951), who reported 4 cases as a complication of lumbar intervertebral disc operations. Davidoff, Gass and Grossman (1947) reported 5 cases of arachnoiditis following operation for benign spinal cord tumors. Their first case had arachnoiditis associated with recurrence of a meningioma fourteen years after the removal of the original tumor. In their fifth case, necropsy disclosed foci of meningiomatous tissue with dense arachnoidal proliferations ten years after the original operation.

It was believed by the earlier authors, notably Stookey (1927) and Elkington (1936), that spinal arachnoiditis was more common in the cervical and thoracic regions. However, French (1946) and Ramsey, French and Strain (1946) indicated that in their experience the occurrence of arachnoiditis in the lumbar region was considerably more common, and this is borne out by my experience. Six of the 7 cases reported by Seaman, Marder and Rosenbaum (1953) also involved the lumbar spinal canal.

The possibility of syphilitic and tuberculous meningitis also must be kept in mind. While now infrequent, it is apt to be overlooked unless its existence is recalled.

The pathologic changes associated with arachnoiditis are characterized by the presence of diffuse adhesions, which may be minimal and localized, or thick, whitish fibrotic intermeshed bands, and occurs in all gradations of severity. Thin, stringy fibrous tissue containing large numbers of polymorphonuclear cells first appear, and later as organization takes place, a thickened membranous fibrotic lesion is formed (Kulowski and Scott, 1934). The arachnoidal inflammation is recurrent and may be diffusely disseminated or locally circumscribed, and associated with the formation of cystic areas. The thickening may be moderate or extensive, and distort the dural sac, extend into the nerve root pouches deforming and compressing the nerve roots themselves (Rexed, 1947). Intramedullary changes, notably cavitation, have been described as a result of pressure and circulatory disturbances (Lubin, 1940).

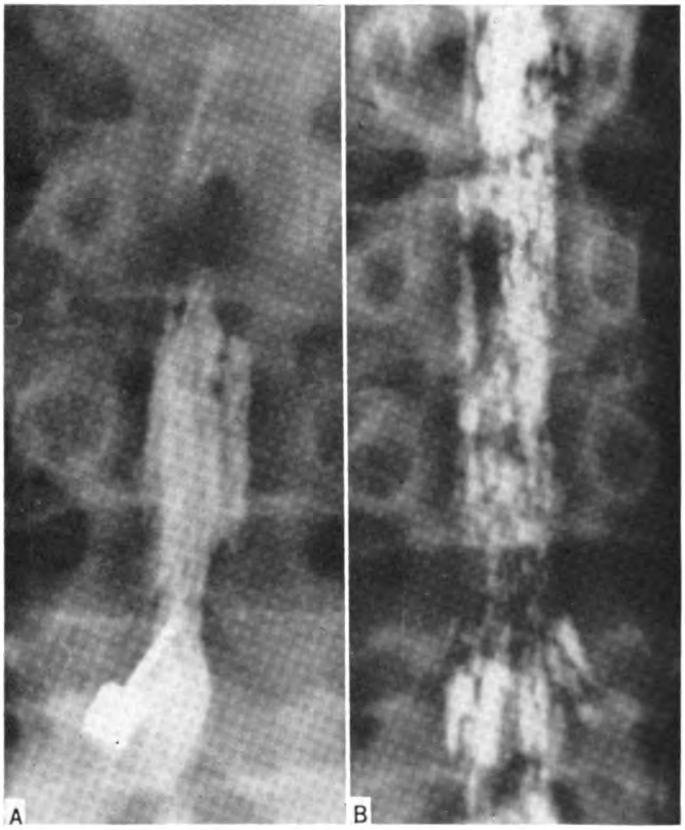

FIG. 326.—Arachnoidits in a 38-year-old man who had had staphylococcic meningities 20 years before, treated with multiple lumbar punctures and intrathecal medication including prontosil, sulphapyridine and bacteriophage. The flow of cerebrospinal fluid was rather slow, and 3 cc. of pantopaque was instilled (*A*). Under fluoroscopic inspection the flow was very slow, and the column broke up into many droplets which did not reunite (*B*).

Examination of the cerebrospinal fluid may be of help. It should be kept in mind that manometric readings may range from normal values to complete block. The cerebrospinal fluid protein content may vary from normal to an extremely high value with xanthochromic or thick yellowish fluid. French (1947) mentioned that the protein value may exceed 400 mgm. per cent, and we have observed even higher contents.

Over the past twenty years we have seen 27 cases of arachnoiditis. Only 2 of these involved the dorsal spinal canal, and the clinical picture in both patients was that of a spinal cord tumor, with complete block in the mid-dorsal area. The true nature of the lesion was disclosed at operation, and the postoperative results were poor. In 8 cases arachnoiditis involving the lumbar spinal canal was concomitant with large herniated discs. In all the dura was opened to gain access to the prolapsed discs, and the thickened nerve roots and arachnoidal inflammations were then identified.

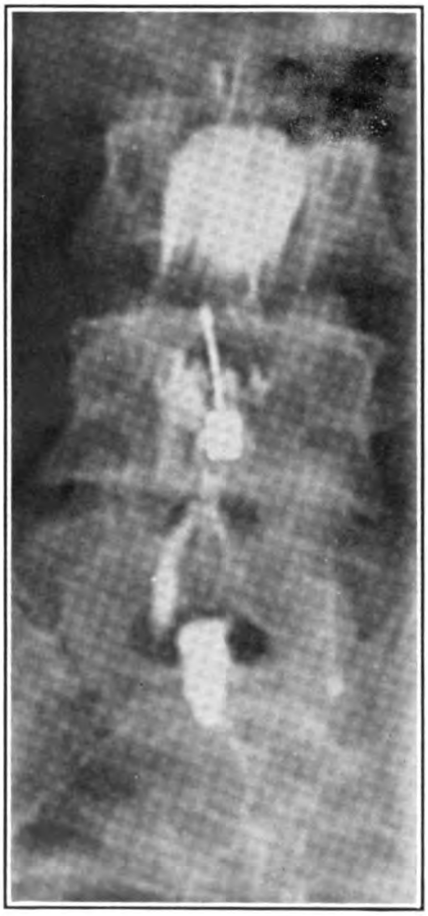

FIG. 327.—Arachnoiditis following removal of herniated disc from fourth lumbar interspace. Recurrent pain, with almost complete block at the third lumbar interspace. Irregular collections of pantopaque below the obstruction were caught in thick, fibrinous exudate which had almost obliterated the spinal canal.

These cases appear to fall into a group described by Kennedy, Elsberg and Lambert (1924), who depicted radiculitis of the cauda equina as a separate entity in which there was relatively little evidence of inflammatory reaction other than thickened nerve roots. It may be significant that in all of these cases exploration of the spinal canal by the lateral approach failed to disclose the protruded discs, which were identified only after the dura had been opened. Almost all had complete or almost complete spinal canal block.

In 12 cases arachnoiditis was associated with previous surgery for herniated discs. In 5 of these a second operation disclosed in addition to the arachnoiditis, recurrences of the protruded discs. The time interval between the first operation and the need for subsequent operation was variable. In 1 case only six weeks elapsed, while in the others the intervals varied from three to ten years. One patient had had intrathecal medication for meningitis about ten years before, and another had had a briskly bleeding aneurysm of the circle of Willis six months before the appearance of spinal symptoms.

In 5 other cases there was no known reason for the arachnoiditis, and in each case the diagnosis was either spinal cord tumor or herniated disc.

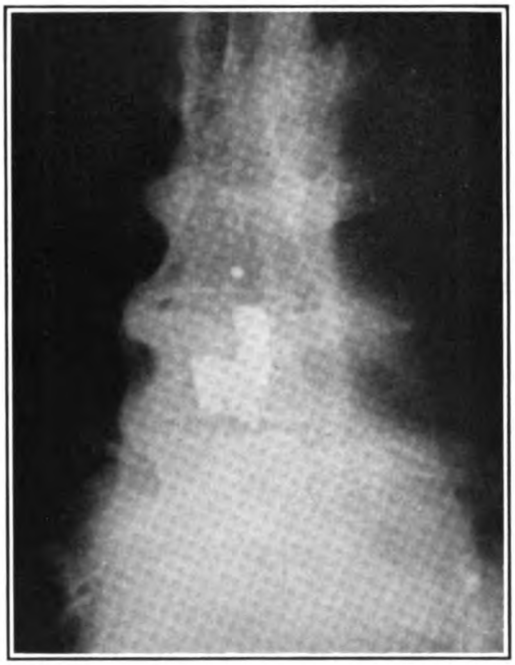

FIG. 328.—Block at the level of the seventh thoracic vertebra due to arachnoiditis of unknown origin. The clinical syndrome was that of a spinal cord tumor. Operative verification.

The plain roentgenograms in our cases of arachnoiditis, as well as in those reported by others, were usually normal. When no known cause can be postulated, the demonstration of a previous laminectomy may be a helpful hint. The diagnosis of arachnoiditis is best made by means of myelography, and the picture is indeed a variable one. In its most characteristic form myelograms reveal a spotty irregular, loculated pattern which may extend over several segments. When this exists the passage of pantopaque through the area is slow and irregular, but no single point of abrupt obstruction is observed. In a simpler form, and particularly when associated with large protruded intervertebral discs, radiolucent streaks parallel to the long axis of the canal may be observed. These terminate in a dentate pattern at the line of the block and correspond to the thickened roots of the cauda equina. Associated with this may be irregular defects in the pantopaque column caused by patches of agglutinated tissue. In such cases, when no other disruption of the pantopaque column is present, a spinal cord tumor may be simulated.

The most variable patterns are encountered in the lumbar or lower thoracic areas. French (1947) mentioned that in cases of recurrent arachnoiditis of the dorsal spinal canal myelography may disclose an irregular appearance of the oil column with partial or complete block. Lubin (1940), in a report of adhesive spinal canal arachnoiditis involving the cervical areas, included a thirty-six-year-old man in whom combined cisternal and lumbar punctures revealed a complete block. At necropsy this patient had intradural adhesions extending from the third to the sixth cervical segments. The arachnoid was markedly hyperplastic and adherent to the cord. Cavitations within the cord were attributed to interference with its blood supply. Blumstein and Baker (1943) believed that arrest of iodized

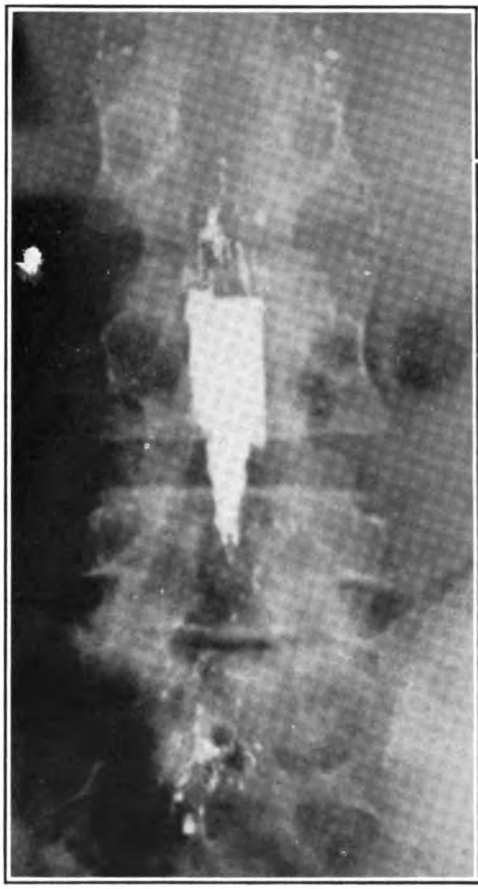

FIG. 329

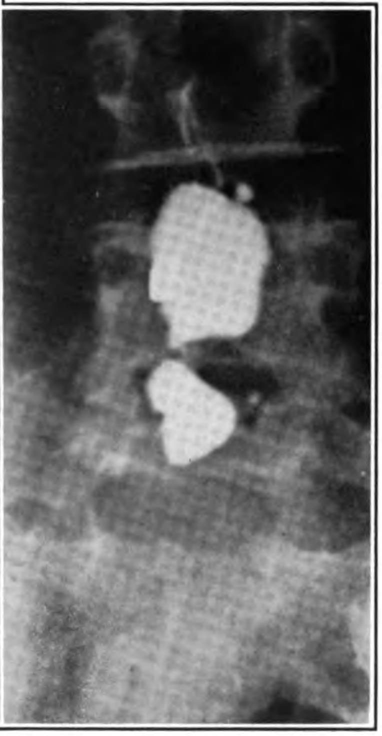

FIG. 330

FIG. 329.—Post-operative arachnoiditis involving lower lumbar spinal canal, erect myelogram. Note the tapering of the caudal end of the pantopaque column and block at the level of the laminectomy. Operative verification.

FIG. 330.—Myelographic defect at the fourth lumbar interspace due to inflammatory adhesions kinking the dural sac against the neural arch and ligamentum flavum.

oil at multiple levels was an important indication of arachnoiditis. Of interest in their report is the description of a case of arachnoiditis with necropsy in a seventeen-month-old infant.

In our cases of lumbar arachnoiditis, particularly those with previous surgery, an interesting change was an irregular tapered thinning of the caudal sac. In some, irregular, small scallops were seen in the lateral margins of the distal thecal sac. The area of narrowing extended for as much as three spinal segments. It was in these cases that recurrent herniated discs could not be identified myelographically. At operation the subarachnoid space in these patients was practically obliterated, but sufficient space remained to permit seepage of the pantopaque downwards. In others a complete block was encountered, and the distal end of the pantopaque column assumed a variable appearance, of which the dentate impressions of the swollen nerve roots was the most frequent. The most frequent pattern was that described above, with slow movement and irregular pooling of the pantopaque. In a few instances the block was complete, and was indistinguishable from a tumor within the spinal canal. The myelograms resembled those described by Lewtas and Dimont, so that I doubt whether the picture is specific for one entity alone.

Clinically, arachnoiditis may simulate spinal cord tumors, multiple sclerosis, radiculomyelitis, Landry's ascending paralysis, or other degenerative diseases of the spinal cord. This again stresses the necessity for myelographic investigations in patients with obscure neurologic pathologic patterns.

Recently I had the opportunity of examining a patient who had had a thorotrast myelogram about 18 years before he came to us. The patient complained of fluctuating pain in both lower extremities, some difficulty in starting the urinary stream, diminished sexual potency and low back pain. On plain film examination of his lumbar spine it was noted that there were thin radiopaque linear markings within the spinal canal which assumed the configuration of the cauda equina. Within his skull the choroid plexuses also were opacified, but were not displaced. It was believed that he might have arachnoiditis following thorotrast examination. Because of this, after much discussion as to whether or not myelography would contribute to the diagnosis, a pantopaque examination was performed. This showed free flow of the pantopaque from the caudal sac to the cervical spinal canal, with no filling defects. The pantopaque was removed from the spinal canal without difficulty. It became obvious that no arachnoiditis of clinical significance existed, and that surgery was not required. Because of the impregnation of the choroid plexus with thorotrast one could conclude that a similar process had resulted in opacification of the filaments of the cauda equina. A review of his past history disclosed that the complaints that brought him to us were quite similar to those he had had prior to the thorotrast myelography.

Epidural Adhesions

We have observed 2 cases in which the dura at the level of the fourth and fifth lumbar intervertebral spaces was sharply kinked and adherent to the overlying ligamentum flavum. The adhesions also extended under the neural arches. In 1 of the patients, a twenty-eight-year-old woman, the clinical diagnosis was herniated disc, and myelographic examination disclosed a semicircular filling defect on the left side of the canal at the fourth lumbar interspace, which was considerably narrowed. At operation no disc protrusion was encountered and only kinking of the dural sac was observed. Before the adhesions were separated no pulsations were observed in the spinal epidural vessels below the area of adhesions. However, when the adhesions were separated pulsations returned. No reason for the presence of this lesion could be ascertained.

In the second case, the patient had had a previous operation at which a large protruded disc had been removed from the fourth lumbar interspace and a smaller one from the lumbosacral interspace. Another myelogram three years later revealed an obstructive defect at the interspace between the fourth and fifth lumbar vertebræ, and it was believed that a recurrent herniated disc existed. There was a free flow of pantopaque through the entire spinal canal above this level. At operation, adhesions were encountered between the dura, the ligamentum flavum and the adjacent bone, causing the defect noted on the myelograms.

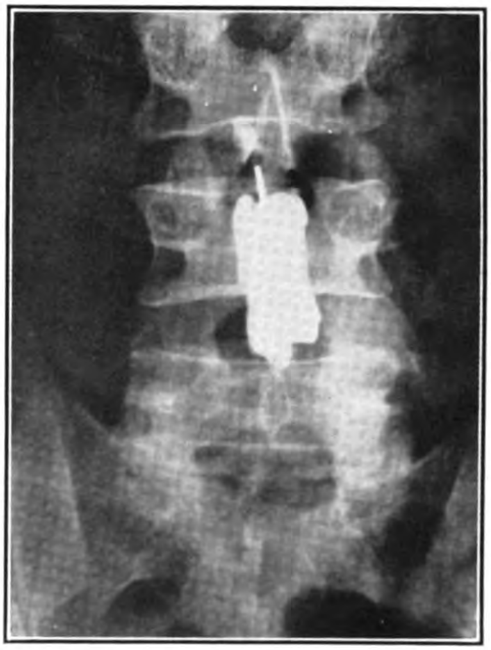

Fig. 331.—Obstruction at the fourth lumbar interspace due to epidural scarring following removal of herniated disc three years before recurrence of back and sciatic pain. At operation the dural sac was opened and arachnoiditis involving the cauda equina also was present.

The latter case is much the same as those described by Knuttson (1949), who mentioned that the deformity may be due to postoperative hemorrhage with thickened scar formation and adhesions. In such cases the impression of a recurrent disc may be difficult to refute, and he reported 3 cases in which a second operation showed no recurrence of a herniated disc but only scar formation. He concluded that the deformity followed extirpation of the ligamentum flavum followed by adhesions, together with a local hernia-like protrusion of the posterolateral aspect of the dura.

Epidural and Subdural Infections

Epidural infections usually are situated in the dorsal aspect of the spinal canal, and may extend laterally to some degree. The reason for this lies in the fact that the epidural space between the dura and the periosteum is widest in this area. Anatomically the dura is so situated that its ventral aspect is in close approximation to the vertebral bodies and the intervertebral discs, rendering the epidural space quite narrow anteriorly. The epidural space is widest in the thoracic and lumbar regions, and is minimal in the cervical area. Below the seventh cervical segment the epidural space may reach a depth of 0.5 to 0.7 cm. Below the fourth to eighth dorsal vertebræ it again tapers and becomes shallow below the eleventh dorsal to the second lumbar vertebra. Over the remaining lumbar vertebræ the epidural space attains its greatest depth. Within the epidural space is a layer of fat, connective tissue and arterial and venous plexuses (Reeves, 1940). It is within the fatty and loose areolar tissue that infection may take place. Such infectious processes may be either acute, subacute or chronic. The most common cause is metastatic abscess formation secondary to a staphylococcic infection elsewhere in the body. According to Grant (1945) the presence of free pus in the epidural space without granulation tissue on the dura constitutes the acute phase of an epidural abscess. If both free pus and dural granulations are found the condition is considered subacute, and if a solid epidural fibrous mass without definite macroscopic evidence of infection is discovered the abscess is considered of a chronic nature. In neglected lesions the pus may track into the paravertebral muscles as well as the epidural space (Echols, 1941). Browder and Meyers (1937, 1941) considered that all such infections were preceded by vertebral osteomyelitis, and that the extension of the infection was from the bony structures to the epidural space. Rankin and Flotow (1946) commented that epidural infections almost never resulted from leptomeningitis perforating the dura. Raney (1939) reported 3 cases of pneumococcic epidural abscess associated with pneumonia. Rangell and Glassman (1945) described a case which they believed to be the first of this variety in which an acute spinal epidural abscess followed a diagnostic lumbar puncture within three days.

Negrin and Clark (1952) also reported that pyogenic infections may take place by direct contaminations of the subdural space by a spinal or caudal needle puncture. They differentiated this from metastatic epidural infections by the fact that the latter overlay the cord, at least in part, while subdural extension involves particularly the region of the cauda equina. In either event surgical intervention is required for drainage of the purulent contents practically as an emergency procedure.

Another route for epidural infections is by way of congenital dermal sinuses. Such cases were described by Walker and Bucy (1934). Stammers (1938) also reported 2 cases of interest in children, 1 a two-year-old child with a low grade meningitis due to a dermal sinus which extended from the surface of the upper sacral region through a bifid fifth lumbar vertebra to communicate with a dermoid cyst within the spinal canal. The second case, a four-year-old boy, also had meningitis incident to a discharging sacrococcygeal sinus attached to the dura. Waring and Pratt-Thomas (1945) also reported 2 cases in which a congenital dermal sinus was a source of meningeal infection, the sinus extending from the surface in the lumbar region down through a defect in the subjacent vertebra communicating with the dura and giving rise to an epidural abscess. Mount (1949) mentioned a case of congenital dermal sinus in the upper thoracic region which led to an abscess within the spinal cord in a ten-year-old girl.

Spinal epidural abscesses also may be associated with streptococcus in fections, and according to Krumdieck and Stevenson (1940) infections with actinomycosis as well as with typhoid bacillus and bacillus pyocyaneus have also been reported in isolated cases. Heusner (1948) collected 200 cases of spinal epidural sepsis.

Epidural granulomas may form either as a result of chronic pyogenic infections, or may be associated with tuberculosis, syphilis, actinomycosis, or coccidiodal infections. They may also be non-infectious in nature, such as foreign bodies (Bucy and Oberhill, 1950). These are mass lesions, and although of similar etiologic origin, are considered under the heading of tumors. Spinal epidural hematomas result in similar changes and are considered later in this chapter.

The clinical picture of spinal epidural infections varies with the extent of spinal cord compression as well as the degree of toxicity produced by the process. In acute cases a rapid progression of pain and paralysis below the level of involvement takes place, together with local tenderness and fever. Sphincteric disturbances and motor weakness may occur within a week after the onset of pain. The pain may be accentuated by bending or other movement, and associated with nuchal rigidity, a positive Kernig sign and muscular spasm. In subacute and chronic cases symptoms may be slower in progression, and the total effect may simulate an intraspinal tumor.

As a rule the plain film roentgenograms of the spine are not helpful. However, in all cases suspected of an epidural infection, a very careful study of the vertebral structures for breakdown of bone pointing towards osteomyelitis should be undertaken. In such cases stereoroentgenograms are helpful, and in doubtful instances the use of body section radiography for detailed inspection of the vertebral bodies and the neural arches provides diagnostic information. One should not hesitate to employ myelography in patients suspected of harboring an epidural abscess, particularly of the acute variety. In such cases the myelogram may disclose a complete block at the level of the lesion, and if this or any other filling defect is noted, immediate surgical intervention may be indicated. In some instances it is advisable to instill the pantopaque both by the lumbar and the cisternal routes, particularly in those cases in which a complete block is present and symptoms are referable to a wide area of the spinal cord. In 1 case encountered by us the epidural infection extended from the fifth dorsal to the second lumbar vertebræ, and was associated with an osteomyelitis of the body of the first lumbar vertebra.

Freedman and Alpers (1948) observed that the occurrence of spinal subdural abscesses was rarely mentioned in the literature. They reported 2 such cases, with signs and symptoms characteristic of an intraspinal abscess, namely, back pain which occurred with an acute inflammatory disease, followed by signs of progressive involvement of the spinal cord, and accompanied by subarachnoid block. In their 2 cases there was no evidence of extradural inflammation or deformity of the cord. The dura was thickened and infiltrated with inflammatory cells, with the formation of subdural abscesses. In 1 case the inflammation extended from the cervical region to the cauda equina. The authors noted that the preoperative differentiation of extradural and subdural spinal abscess is probably impossible, an observation reiterated by Negrin and Clark (1952).

Hypertrophic spinal pachymeningitis is described by Bucy and Freeman (1952) as a disease of unknown etiology which results in thickening of the dura with proliferation of collagenous fibers arranged in parallel rows and

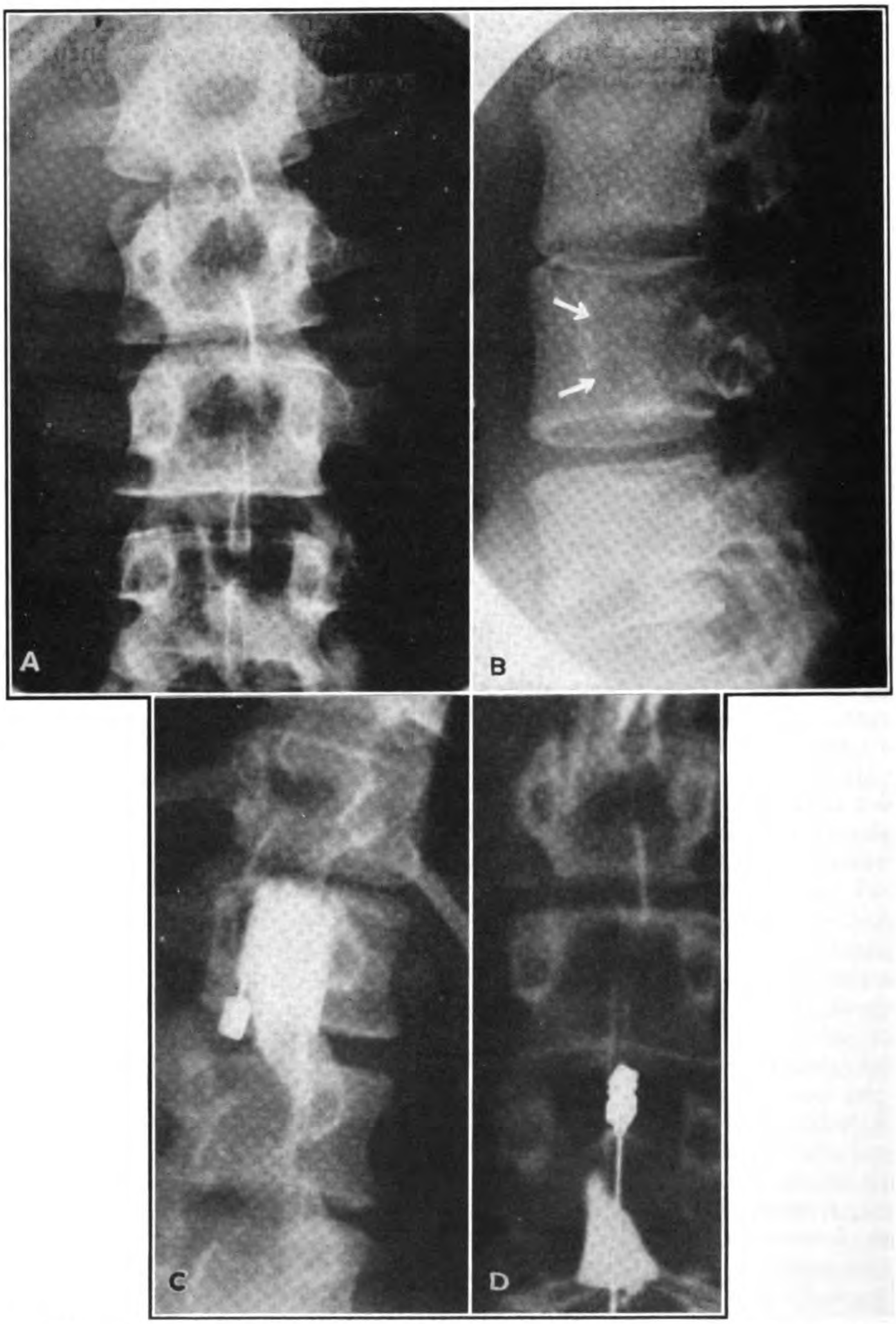

FIG. 332.—*A*, Anteroposterior view of lumbar spine of eighteen-year-old male patient with rapidly progressing weakness and paraplegia of lower extremities. No definite osseous changes in this projection. *B*, Same case, lateral view. Note the destruction of bone in the posterior aspect of the body of the second lumbar vertebra (arrows). *C*, Pantopaque instilled at the first lumbar interspace. Note the block at the second lumbar interspace. *D*, Same case. Pantopaque instilled at lumbosacral interspace, with block at the fourth lumbar interspace. At necropsy the epidural space between the upper and lower obstructions was obliterated by an abscess emanating from the second lumbar vertebra. It could not be ascertained whether this was purely osteomyelitis or a degenerated neoplasm.

infiltrated with cells characteristic of a chronic inflammatory reaction. This occurs most often in the cervical spinal canal, but may occur elsewhere as well. In the 2 cases reported by these authors 1 occurred in the lower cervical spinal canal and the other in the upper dorsal spinal canal. The clinical picture was one of radicular pain, muscular weakness and atrophy similarly localized, associated with spasticity and muscular weakness below the level of the cord compression. The function of the bowel and bladder may be impaired frequently. Plain roentgenograms in this condition are normal, but myelograms will reveal evidence of block, thus localizing the level of the lesion.

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INTRASPINAL DERMOIDS AND EPIDERMOIDS, AND DERMAL SINUSES

Hamartomas are heterotopic formations composed of elements of the skin, and are dysembryonic malformations rather than true neoplasms. They are regarded as tumors because of their expanding nature. Epidermoids consist of tissues derived from the superficial layers of the skin. Dermoids originate from all layers of the skin, both epithelial and mesenchymal, with accessory cutaneous organs such as sebaceous glands, sweat glands and hair follicles. Dermal sinuses are tubular formations which may have the histologic characteristics of epidermoids, dermoids or both, but which are always in direct communication with the skin. In all three conditions the viable cells form a thin capsule, and the interior is composed of cheesy amorphous masses of dead cornified epithelial cells. In the case of dermoids, sebaceous matter and hair are encountered. List (1941) classified hair-containing tumors as dermoids in his study, even though microscopic examination of the cyst wall showed no cutaneous structures.

Epidermoids occur in a fairly uniform distribution over the thoracic and lumbosacral portions of the cord, and are most frequent in the region of the cauda equina. Dermoids appear to have an affinity for the lumbosacral portion of the cord. Dermal sinuses occur chiefly in the lowermost portion of the spine or in the upper thoracic region. They are seldom encountered in the cervical region. As a rule, these lesions are single, but instances of multiple epidermoids or dermoids, either due primarily to multiple origin or to implantation metastases in the subarachnoid space from rupture of the main nodule have been recorded.

Communications between pilonidal sinuses and intraspinal dermoids were described by Sachs and Horrax (1949). These authors collected 14 such cases from the literature, and considered them due to congenital causes, probably simple mechanical defects in fusion. They were able to assemble 59 dermoid and epidermoid tumors of the spinal cord from the literature, to which they added 2 cases. There were 25 teratomas of the spinal cord reported, and of this group the dural or intradural tumor was associated with a congenital dermal sinus in 22 cases. They reported the case of an adult male with slowly progressive neurologic symptoms in which a congenital dermal sinus in the cervical region communicated with the dura and an intraspinal dermoid.

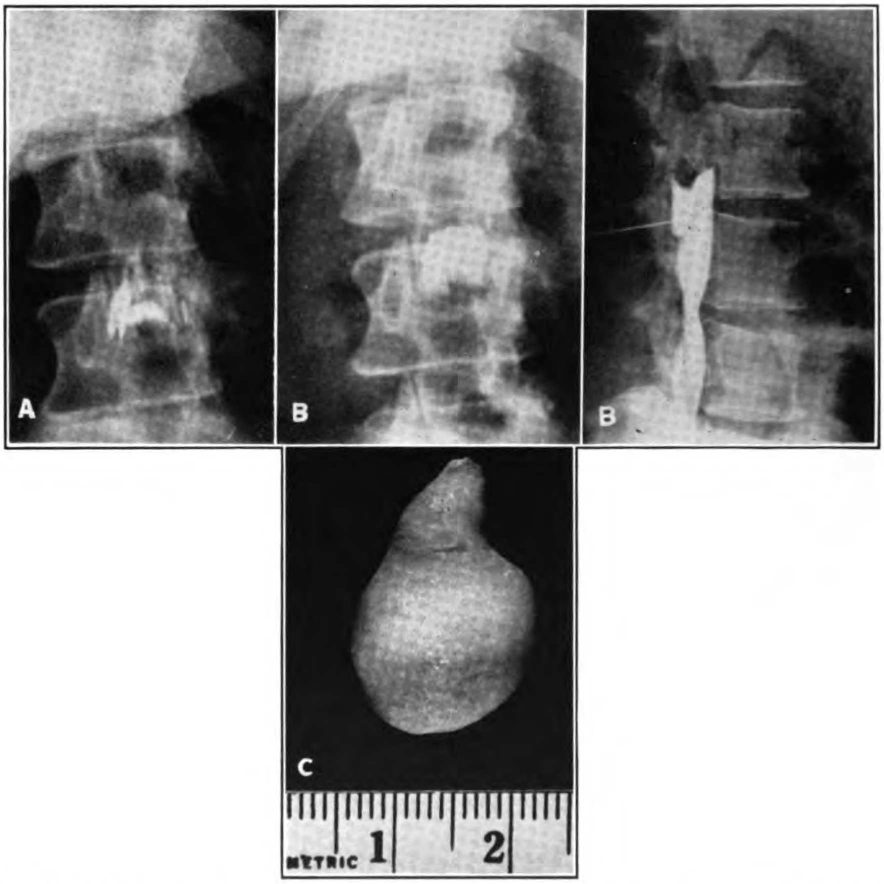

FIG. 333.—*A*, Block at the level of the second lumbar vertebra, superior aspect. Cisternal instillation of pantopaque. *B*, Same case. Pantopaque instilled from below reveals an arcuate block at the level of the pedicles of the second lumbar vertebra. *C*, Same case, operative specimen, epidermoid, epidural in position.

Sacrococcygeal teratomas in infants and children were reviewed by Ravich and Smith (1951), who described 9 verified cases of their own. Large tumors which presented externally, were present at birth but sometimes remained inconspicuous until rapid growth and malignant changes appeared. Ulceration and infection may lead to the death of a patient while the tumors are still benign. Obstructive symptoms referable to the rectum, the ureters or urethra appear because of extrinsic pressure.

Malignant transformation of these growths is relatively common, the usual type being papillary adenocarcinoma.

In 3 of the cases reported by Ravich and Smith (1951) death occurred because of metastases, and in 1 case there was a massive local recurrence after incomplete removal of a benign teratoma.

Intraspinal epidermoid tumors also occur in adults. Moore and Walker (1951) reported 1 in a fifty-two-year-old man who had had symptoms over twenty years. Malignant degeneration in an intramedullary teratoma was reported by Lemmen and Wilson (1951) in an eleven-year-old girl. Intraspinal epidermoids may be either intradural or extradural. According to Craig (1943), who gathered 43 such cases, half of the 20 cases of epidermoids were intramedullary. Of the 23 dermoids, an extramedullary situation was more frequent, and these were occasionally extradural. He reported a subpial epidermoid associated with spina bifida of the first sacral segment. Occasionally a teratoma may arise within the spinal cord and extend to the extramedullary region. Such a case was reported by Black and German (1950) in which the tumor practically filled the entire spinal canal. Mosberg (1951) commented upon the high association of intraspinal tumors with other congenital anomalies during the first year of life.

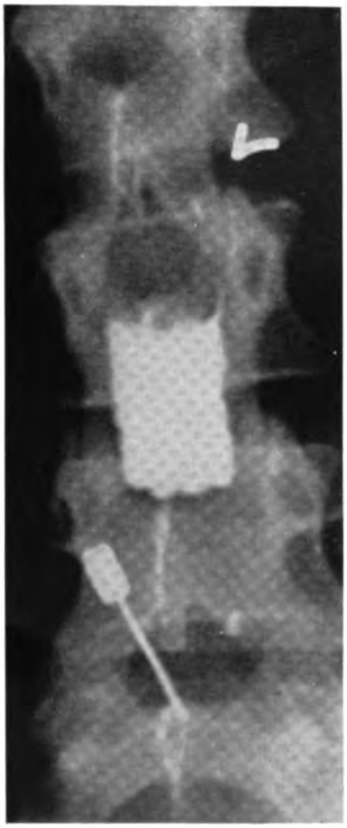
FIG. 334.—Complete block due to extradural dermoid at the level of the third lumbar vertebra in a nineteen-year-old girl, who had been considered "neurotic" for two years.

Plain film roentgenograms with congenital large tumors reveal widening of the anteroposterior and lateral diameters of the spinal canal, with erosion of the pedicles and sometimes arcuate indentations in the dorsal aspects of the adjacent vertebræ. Spina bifida is frequently present. These changes sometimes are sufficient for diagnosis, and myelography becomes unnecessary. However, if the plain films are normal, as is more frequent in older patients, there should be no hesitation to employ myelography, particularly if a dimple is present in the mid-line overlying the suspected area. As a rule myelograms in such cases will show only block rather than any characteristic defect, except perhaps for the existence of diastematomyelia (Naffziger and Jones, 1935; Ingraham and Bailey, 1946; Hannan and Geist, 1950). In occasional cases the head of the pantopaque column assumes a rounded or somewhat scalloped configuration.

One frequently encounters teratomas in the presacral region. According to Mayo, Baker and Smith (1953) congenital tumors accounted for 62 per cent of 161 presacral tumors gathered from the literature. Inflammatory lesions constituted 11 per cent of this series. There were 11 per cent of neurogenic growths, bony neoplasms constituted 9 per cent and miscellaneous growths occurred in 7 per cent. Roentgenologically these tumors manifested themselves predominantly by bone erosion and displacement of adjacent viscera. Operation and pathologic examination are usually required to definitely identify these lesions. McCarty (1950) also observed that the most frequent congenital presacral tumors were dermoids and teratomas, usually attached to the anterior sacral wall and the coccyx.

In a report of 40 cases of sacrococcygeal teratomas in infants and children Gross, Clatworthy and Meeker (1951) noted that the growths were more common in females. Plain roentgenograms in 37 cases showed soft tissue masses at the coccyx in 35. Calcifications were seen in 16 of these patients. Distant metastases to the lungs and bones were observed in 6.

The presence of a cutaneous dimple or of a discharging sinus in the middle of the back suggests the existence of a pilonidal sinus. This in turn may indicate a coexistent intraspinal mass lesion in certain instances. Of equal importance is extension of an infection into the spinal canal by way of a dermal sinus. As a rule these extend to the epidural space and the meninges, but rarely they may enter the intramedullary region. Such a case was reported by Kooistra (1942) in a nineteen-year-old girl with a sinus over the third thoracic vertebra. The plain roentgenograms were normal, but myelograms showed a block at the third thoracic vertebra. The inner aspects of the left pedicles at this level were thinned. The clinical picture was that of a spinal cord tumor, and operation disclosed a mass extending from the seventh cervical to the fourth thoracic vertebra.

Meningitis or myelitis of an acute or chronic nature may follow infection extending along a pilonidal sinus. Craig (1943) suggested that a sterile form of meningitis may be caused by liberation of fatty acids from a cyst within the subarachnoid space. Shenkin, Hunt and Horn (1944) reported a two-year-old infant with a clinical syndrome of transverse myelitis and widespread leptomeningitis caused by infection along a pilondal sinus in direct continuity with a patent filum terminale continuous with the central canal of the spinal cord. When this is suspected it is better to attempt gas myelography before resorting to the instillation of pantopaque, because the injection of foreign material into an infected subarachnoid space should be avoided.

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INTRASPINAL NEOPLASMS

Tumors within the spinal canal occur as intramedullary growths, as extramedullary tumors, or tumors which perforate the dura and are both intradural and extradural but extramedullary. In a review of 557 intraspinal tumors by Rasmussen, Kernohan and Adson (1940) intramedullary tumors constituted 11 per cent of the series. Extradural growths were present in 28 per cent, and combined intradural and extradural growths were present in 8 per cent. Intradural extramedullary growths made up 53 per cent of this series. Eleven years later Woltman, Kernohan, Adson and Craig (1951) reported on 979 intraspinal tumors, and in this group there were 220 intramedullary gliomas, constituting 22.5 per cent of the series. Elsberg (1941) reported that of 275 spinal cord tumors 92 were intramedullary, 175 were extramedullary and 81 were extradural. Of the extramedullary tumors perineurial fibroblastomas were the most common, and meningiomas were almost equally frequent. Both groups together composed approximately 60 per cent of the reported intraspinal neoplasms. These growths occurred either entirely within or without the dura, but the intradural situation was the more frequent. Of the 59 perineurial fibroblastomas reported by Elsberg, 34 were intradural and 25 were extradural. Of his 73 meningiomas, 69 were intradural and 4 extradural as well.

Rasmussen, Kernohan and Adson (1940) reported 67 per cent of 163 neurofibromas as intradural in location and 15 per cent were extradural. The remainder were both intradural and extradural. These figures indicate that the two most frequent growths within the spinal canal are usually intradural, but extradural or both extra- and intradural positions are not uncommon. Among the other extradural growths which may be encountered are sarcomas, hemangioblastomas, chordomas, melanomas, dermoids and metastatic tumors.

Occasionally one encounters multiple tumors within the spinal canal (Lichtenstein, 1941). This is relatively frequent in cases of perineurial fibroblastoma and will be again mentioned under that heading. Occasionally meningiomas may be observed together with perineurial fibroblastomas as an entirely separate but concomitant process. Such a case was reported by Svien, Camp and Adson (1949) in a twenty-nine-year-old woman who had three primary intraspinal tumors. Two of these were intradural meningiomas in the upper thoracic spinal canal, and a neurofibroma was removed from the cervical canal later. Twelve years thereafter the patient had symptoms referable to the cerebellopontine angle and the cervical cord. Myelography revealed a complete block at the fourth thoracic vertebra and a partial block at the first lumbar interspace. Operation disclosed an intradural extramedullary neurofibroma arising from the anterior root of the fifth cervical nerve and another from the posterior root of the right fourth lumbar nerve.

Tumors within the spinal canal, especially meningiomas, are most frequent in the thoracic region, and occur with about equal frequency in the cervical and lumbar areas. Of the 557 cases reported by Rasmussen, Kernohan and Adson (1940) 18 per cent were in the cervical region, 54 per cent in the thoracic region and 21 per cent in the lumbar region. Seven per cent were in the sacral area. Of the 296 cases reported by Elsberg (1941) approximately the same distribution was noted. In the later report by Woltman, Kernohan, Adson and Craig (1951) it was mentioned that of 220 intramedullary gliomas 19 per cent were in the crevical region, 31 per cent in the thoracic region, 48 per cent in the lumbar region, and 2 per cent in the sacral region. This reflected the considerable number of ependymomas which occur most frequently in the lumbar spinal canal.

The clinical picture associated with spinal cord tumors requires no elaboration here. It should be recalled that the classical picture of spinal cord impairment is not constant with tumors, and the symptoms and signs may simulate other diseases such as herniated discs. Toumey, Poppen and Hurley (1950) described such a condition with cauda equina tumors, and reported 48 cases. The coexistence of spinal cord tumors with herniated discs is not unknown, and due care in diagnosis is required to avoid overlooking such a combination of lesions. Atypical syndromes produced by extramedullary spinal cord tumors in the cervical region simulate amyotrophic lateral sclerosis, posterolateral sclerosis and other degenerative conditions of the spinal cord. Symptoms referable to the posterior cranial fossa are produced by high cervical tumors projecting into the foramen magnum. A clinical picture simulating acute anterior poliomyelitis with intraspinal tumors in children was described by Chambers (1952). These patients suddenly became paralyzed and had stiff necks, fever and high spinal fluid cell counts. In 1 such case myelography revealed a block, and 2 others had destructive bony changes. One case was verified as an angioma.

Exaggeration of the usual lumbar lordotic curvature in children was mentioned by Furlow (1950) as a sign of cauda equina tumor. One of his 2 cases had a large epidermoid tumor and the second had a neurofibroma. The need for myelography was stressed when there was question as to the cause of an exaggerated lordosis. Craig, Svien, Dodge and Camp (1952) regarded night pain, marked scoliosis or kyphoscoliosis and a known tumor elsewhere as reasons for venturing the diagnosis of spinal cord tumor in patients suspected of herniated discs. These authors emphasized that a considerable elevation of the cerebrospinal fluid protein content was in favor of an intraspinal tumor. However, this is not entirely reliable.

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INTRAMEDULLARY TUMORS

These tumors are predominantly gliomas. In a series of 35 cases reported by Woods and Pimenta (1944), 12 were ependymomas, 7 astrocytomas, 4 spongioblastomas, 2 glioblastomas, 2 mixed gliomas and 1 was an oligodendroglioma. Of the others, 2 were unclassifiable, 1 was a melano-

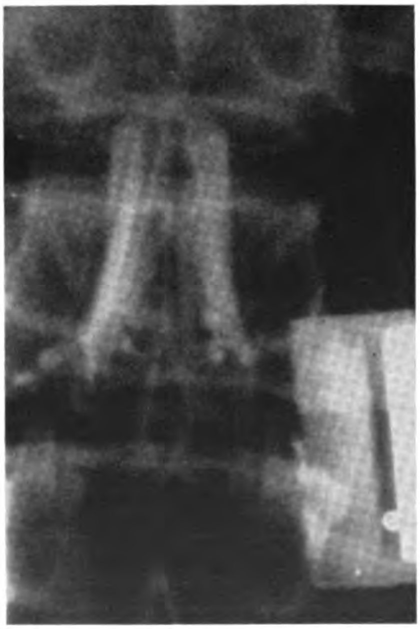

Fig. 335.—Intramedullary glioma verified by inspection, cisternal injection of pantopaque. The distal portion of the contrast medium is deflected laterally on either side by the intramedullary growth.

blastoma, 3 were dermoid cysts and 1 patient had an intraspinal blood clot. They mentioned that they found references in the literature to 7 cases diagnosed as intramedullary tumors, and which at operation were intramedullary tuberculomas. Intramedullary metastatic tumors are rare.

Because of their intramedullary position, these tumors do not produce changes in the bony walls of the spinal canal unless they achieve sufficient size to compress the adjacent pedicles, as for example with the large ependymomas involving the cauda equina. In some instances ependymomas affect the pedicles, laminæ or the anterior bony walls of the spinal canal for one or two segments, but with more bulky tumors almost the entire lumbar spinal canal may be involved. These lesions sometimes assume sufficient size to atrophy the neural arches, spread and thin out the pedicles so that they are almost invisible, and indent the dorsal aspects of the vertebræ producing multiple scallops in this region. Ependymomas originate most often in the filum terminale, and occasionally extend up into the lower thoracic spinal canal. Horrax and Henderson (1939) mentioned an epen-

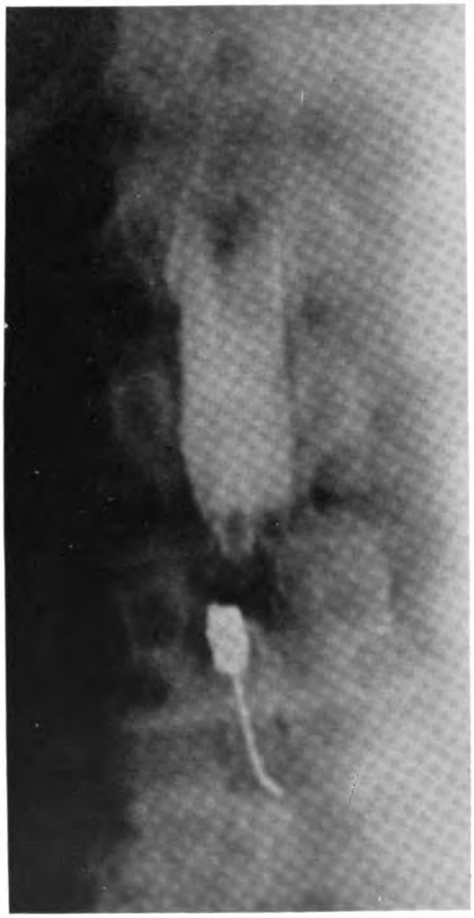

Fig. 336.—Complete block at level of first lumbar vertebra due to verified intramedullary glioma. The lateral extensions of pantopaque are not spread in this case because the lesion was situated in the tapered distal tip of the spinal cord.

dymoma extending from the foramen magnum to the third lumbar vertebra, the tumor measuring 38.5 cm. in length.

Myelography is important in the diagnosis of intramedullary tumors other than large ependymomas. The typical picture produced by gliomas is spinal canal block. The head of the column assumes a cap-like configuration on lumbar myelography, and extending cephalad from this rather shallow deformity one may identify streaks of contrast medium to either side of the swollen spinal cord. If the tumor is sufficiently large, none of the pantopaque will go beyond the lesion. These lateral extensions may present a beaded appearance, which as noted by Walker, Jessica and Marcovich (1941) represent contrast medium in the nerve root sheaths. While the picture of a concave filling defect with upward extending streaks embracing a swollen area within the spinal cord is the customary picture of an intramedullary growth, it is not infrequent to observe a block as the sole myelographic indication. Atypical configurations of the pantopaque column with irregular margins of the lateral aspects or transverse defects also occur. If it is necessary to identify the superior aspect of the tumor in the presence of complete cerebrospinal fluid block, pantopaque is instilled cisternally and the examination continued with the patient erect. No more

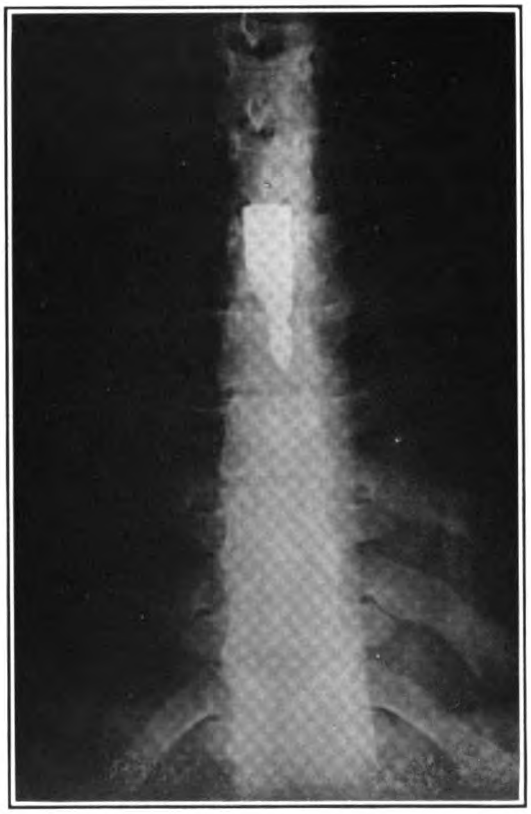

FIG. 337.—Complete block of an atypical configuration in the midthoracic spine, with cisternal injection of pantopaque. Intramedullary glioma found at operation.

than 0.5 to 1.0 c.cm. of pantopaque is instilled if block is suspected even if the flow of fluid is free and manometric determinations are normal or equivocal. If block is present it is preferable not to remove cerebrospinal fluid, and only that pantopaque which can be withdrawn easily is evacuated.

Other spinal cord lesions may be associated with bulging of the cord or obstructions within the spinal canal, and produce a similar picture. Among these are large herniated cervical discs or ridges and syringomyelia. Syringomyelia as a rule is associated with widening of the spinal cord, usually in the cervical area. This produces a widening of the radiolucent space corresponding to the spinal cord, with relatively narrow lateral channels of pantopaque. Myelographic evidence of block consists of hesitation in the passage of pantopaque at the inferior margin of the distended spinal cord and rather slow passage of the contrast material to either side. Intramedullary blood clots, tuberculomas and lipomas are among other conditions which may have to be considered. While large ependymomas rarely require myelographic confirmation, there have been several instances in our experience in which ependymomas were small and the plain film roentgenograms were normal. Myelograms revealed an irregular distribution of pantopaque usually accompanied by partial or complete block. Shenkin and Alpers (1944) noted that infiltrating gliomas may assume many shapes which involve the entire circumference of the cord at one level, taper off at others and invade only a small portion of the cord at several levels. They differentiated between intramedullary ependymomas occuring above

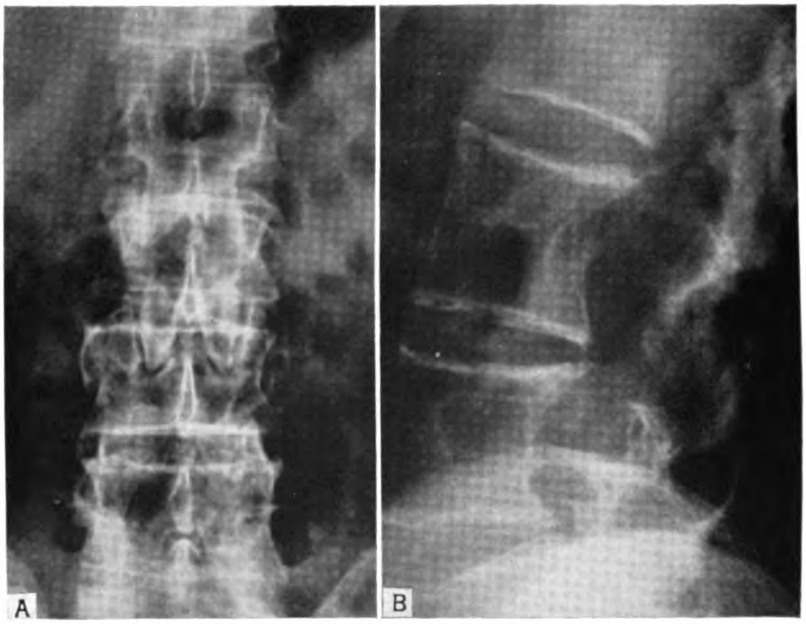

FIG. 338.—Ependymoma of lumbar spinal canal in a 62-year-old man with low back pain radiating down right leg, weakness of right lower extremity, foot drop, and diminished sensation. The pedicles on the left side are destroyed, and the neural arches of the midlumbar vertebræ are thinned (*A*). The lateral projection (*B*) clearly shows the destruction of the pedicles and of the posterior aspects of the second and third lumbar vertebræ.

the level of the filum terminale which were usually of moderate size and the more bulky tumors beginning in or adjacent to the filum. When the tumors were small as with intramedullary ependymomas, bony changes were absent but myelography revealed block.

In a discussion of gliomas of the spinal canal Cooper, Craig and Kernohan (1951) commented on primary extramedullary gliomas. They reported 15 such tumors along the spinal axis which apparently had no attachment to and had not arisen from seeding from a primary intramedullary or intracerebral neoplasm. Included in this series were 9 cases in which the growths were intra- and extradural. The first 9 cases mentioned presented myelographic evidences of block. These authors believed that the heterotopic gliomas arose from glial tissues which had been pinched off from the neuraxis in the course of its development. These unusual growths may be associated with other anomalies, the most frequent of which is spina bifida. Other abnormalities include club feet, undescended testes and spondylolisthesis. Prior to this communication Bailey (1936) had reported a case of astrocytoma of the leptomeninges. He too noted that the glial heterotopias were often accompanied by other anomalies. While it is usual to find that intramedullary growths are inoperable, not every case is necessarily hopeless. Grant

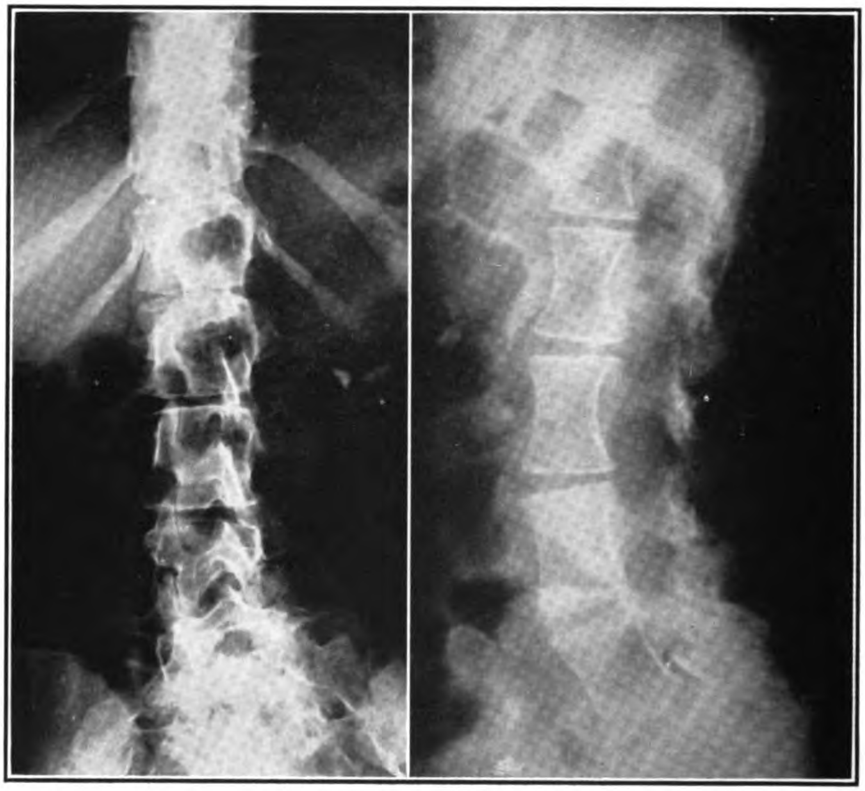

FIG. 339.—Ependymoma of the lumbar spinal canal. This was a huge tumor which filled the spinal canal, and produced marked compressive changes in the dorsal aspects of the lumbar vertebræ. This configuration is sometimes referred to as a "dog spine."

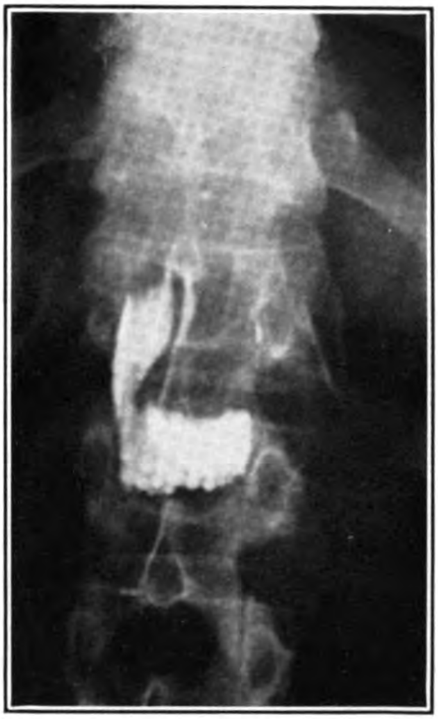

FIG. 340.—Ependymoma at the level of the interspace between the twelfth thoracic and first lumbar vertebræ, with complete obstruction.

(1944) reported the successful removal of a large intramedullary encapsulated astrocytoma from the upper thoracic region of an eighteen-year-old male.

Localized expansion in the spinal canal has been observed in the absence of tumor as a congenital abnormality. Jefferson (1955) reported two such cases in which exploration of the thoracolumbar junction disclosed a normal spinal cord in an enlarged subarachnoid space.

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INTRASPINAL LIPOMAS

These tumors constitute approximately 1 per cent of all intraspinal growths and appear either as intradural or extradural masses. Frequently they attain considerable size and manifest themselves by compression of the spinal cord which may start during infancy, puberty or during the third to fifth decades (Taniguchi and Mufson, 1950). As a rule symptoms are slowly progressive. Ehni and Love (1945) reviewed 29 cases of intradural lipomas and found that they occurred chiefly in the cervical and thoracic regions, most often on the dorsal aspect of the cord. These growths are composed of adult adipose tissue which may be accompanied by an excessive amount of fibrous stroma. There is a tendency, particularly in infancy, for such tumors to be accompanied by congenital deformities, particularly spina bifida. Wycis (1953) reported a case in a forty-eight-year-old woman who also had a Klippel-Feil deformity. In this instance the lipoma was of considerable size and had practically replaced the spinal cord tissue. Wycis postulated that the Klippel-Feil deformity might be

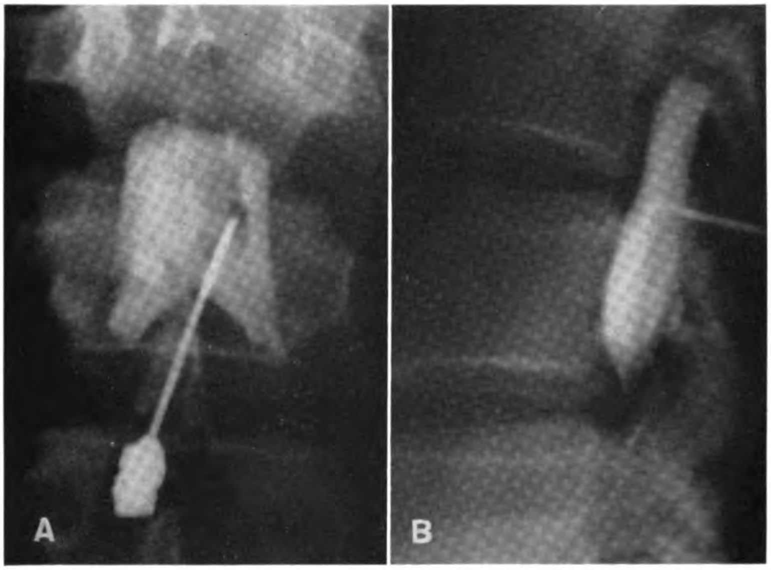

FIG. 341.—Intradural lipoma involving the conus in the upper lumbar region in a 41-year-old man with a subcutaneous lipoma over the midline lumbosacral area. No pain or motor loss. Diminished sensation in sacrococcygeal region, penis and scrotum. Constipated, but patient had difficulty in voiding and catheterized himself for relief. No loss of potency. The lumbar taps in the lower lumbar interspaces were not productive of a good flow of fluid. At the first interspace a good flow was obtained, and 1.5 cc. of pantopaque was instilled. Note the complete block, and the spreading of the pantopaque column.

one which involved both the skeletal and neurogenic tissues, and that the intramedullary lipoma might have arisen from misplaced cell rests accompanying a myelodysraphia. Of particular interest is the fact that there was a paucity of symptoms even though the tumor was so large that it practically obliterated the spinal cord tissue.

A case of interest was reported by Crosby, Wagner and Nichols (1953), in which a lipoma was situated in an extramedullary but subpial location. When this patient was first operated upon it was believed that the tumor was intramedullary, but close inspection and a second operation disclosed that it was actually extramedullary. The tumor was so enveloped in the spinal cord that its exact location was determined only after the most careful scrutiny. They postulated an extramedullary origin for this growth, and that lipomas may be operable even though at first inspection this seems improbable. Wilson, Bartle and Dean (1940) reported the case of a twenty-one-year-old woman with an intradural spinal lipoma and mentioned that the tumor probably arose beneath the pia arachnoid. It appears that the suggestion that these tumors are actually extramedullary but intrude into the substance of the cord because of close apposition has considerable clinical importance.

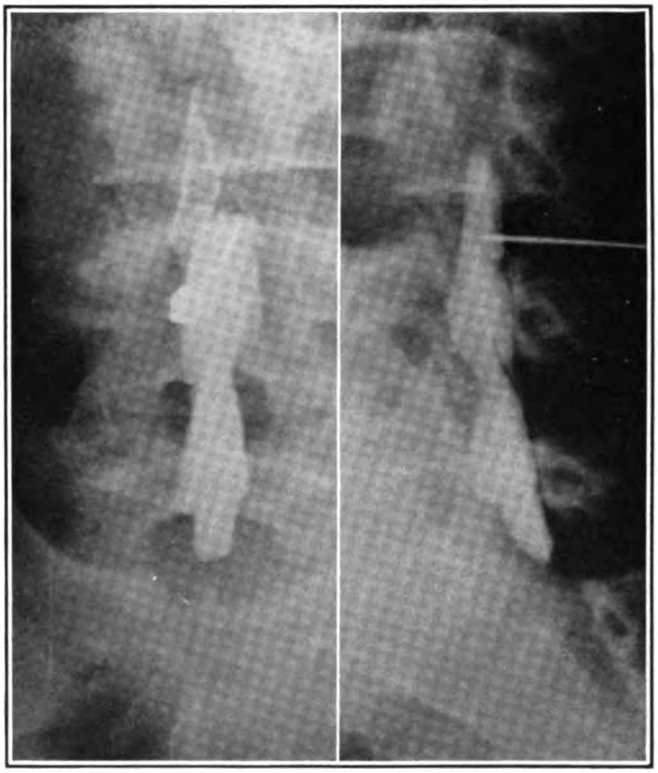

FIG. 342.—Myelographic defect at the fourth lumbar interspace on the left side, anteroposterior and lateral myelograms. The clinical picture in this forty-five-year-old male was characteristic of a herniated disc. At operation an extradural lipofibroma was encountered. Complete relief of symptoms after removal of the tumor.

Lipomas of the spinal cord also occur in an extradural position, and these growths present a somewhat more acute clinical picture. Extradural lipomas are more common in the lower dorsal region, and also occur in the lumbar area and in the cauda equina. Lipomas of the cauda equina usually are associated with other congenital anomalies particularly spina bifida and subcutaneous lipomas overlying the suspected area. In a report of 9 cases Bassett (1950) stated that disturbances of bladder and bowel functions, progressive peripheral neuropathy and trophic changes in the pelvis and skeleton may appear. Bassett believe that lipomas resulted from an overabundance of fatty tissue and protrusion into the dural spaces, or from inclusion within the dura at the time of closure of the neural tube. As the skeletal development of the spine progresses, the conus medullaris comes to lie at progressively higher levels. In these cases the conus may become fixed by the lipoma and the increased traction on the conus and the cauda equina produces symptoms of pressure which may be accentuated by additional accumulations of fat. Ehni and Love (1945) mentioned that only half their extradural lipomas were of adult tissue, most of the remainder being angiomatous in structure, and a few being myxomatous. One was composed of embryonic fat.

Plain film roentgenograms associated with spinal cord and canal lipomas may reveal widening of the canal with spreading and thinning of the pedicles. The presence of other congenital anomalies may suggest the presence of an intraspinal lipoma when neurologic deficiencies are present. The location of the mass can best be identified myelographically. Sometimes, if the growth is of sufficient size, the myelogram simulates that seen with with intramedullary glioma.

Meltzer (1947) reported a lipoma of the lower cervical spinal cord associated with "ivory vertebræ" limited to the neural arches and spinous processes of the sixth and seventh cervical vertebræ.

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INTRADURAL EXTRAMEDULLARY TUMORS

These constitute about 60 per cent of all spinal tumors, and are principally perineurial fibroblastomas or meningiomas. In 275 such cases reported by Elsberg (1941), 159 were intradural, mostly in the cervical and thoracic regions. In a review of 179 cervical intraspinal neoplasms, Webb, Craig and Kernohan (1953) found 68 intradural extramedullary growths. Neurofibromas constituted 38.5 per cent and meningiomas 29.9 per cent. Vascular tumors accounted for 8.4 per cent of their cases. Extradural meningiomas and neurofibromas were also observed. Of the 73 meningiomas reported by Elsberg, 69 were intradural and 4 were extradural. Five of the 59 perineurial fibroblastomas were extradural.

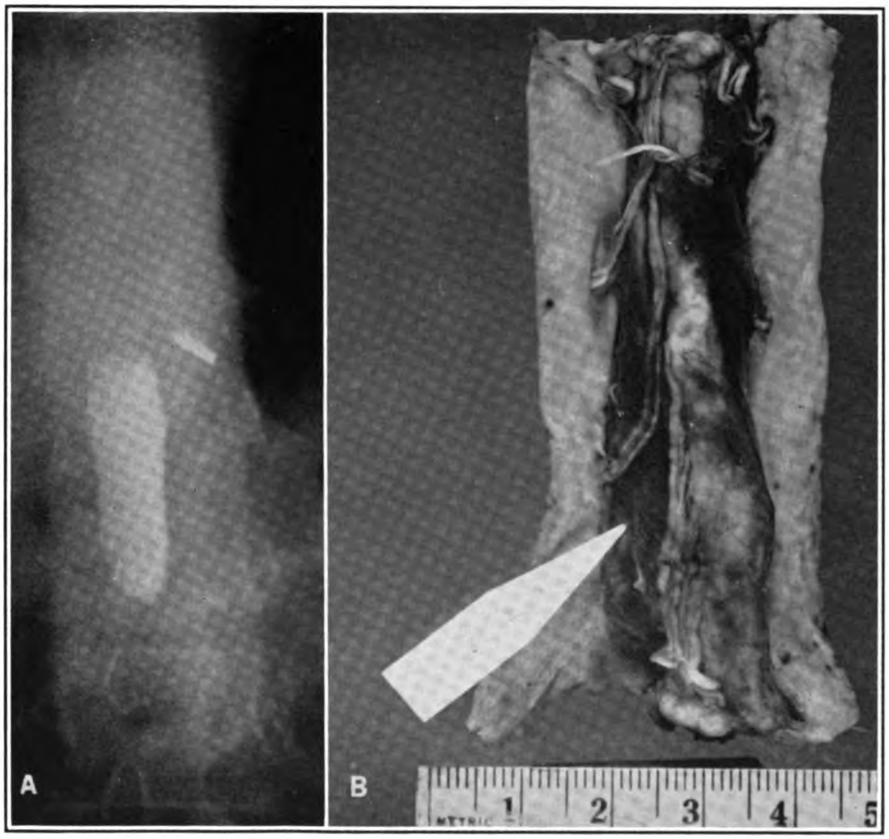

FIG. 343.—*A*, Complete block at the tenth thoracic interspace due to a meningioma. This patient had complained of back pain for two years, and had been paralyzed for several months before myelography was performed. *B*, Photograph of tumor *in situ*, necropsy specimen.

Meningiomas.—These tumors arise from cells of the spinal arachnoid, usually in the villi which lie close to the crossing nerve roots. The arachnoidal attachment of these neoplasms may be minute. Infrequently a meningioma may penetrate the dura and become extradural with disappearance of its arachnoidal pedicle. According to Brown (1942), the pia arachnoid may be incorporated in the tumor, and malignant varieties may cross the pial barrier and actually invade the cord. Of 130 cases of intraspinal meningiomas reported by Brown, 78 per cent were in the thoracic region, 18 per cent in the cervical region, and 4 per cent in the lumbar and lumbosacral regions. There were 8 varieties distributed as follows: meningothelial, 56 per cent; fibroblastic, 28 per cent; psammomatous, 15 per cent; osteoblastic, 4 per cent; lipomatous, 2 per cent; chondromatous and melanomatous, each 1 per cent; and malignant, 6 per cent.

Meningiomas occur more often in females, the incidence varying from 66 to 80 per cent in different series.

A typical intraspinal meningioma occurs as a fairly well-demarcated, smooth or lobulated encapsulated tumor presenting a somewhat spherical or oval configuration. Its capsule is quite tough and broadly attached to the dura. Sometimes it is fixed to the dentate ligament and adjacent arachnoid, with the nerve roots drawn tautly across the tumor. Occasionally no dural attachment can be identified. The tumor seldom invades the extradural fat or adjacent bone, and dumbbell configurations are observed only rarely. Meningiomas extend over one, two or sometimes three interspaces, and are usually dorsolateral in position. Occasionally they occur ventrolaterally or encircle the spinal cord. On cut section they are fleshy, firm and relatively avascular, although the surrounding tissues may become

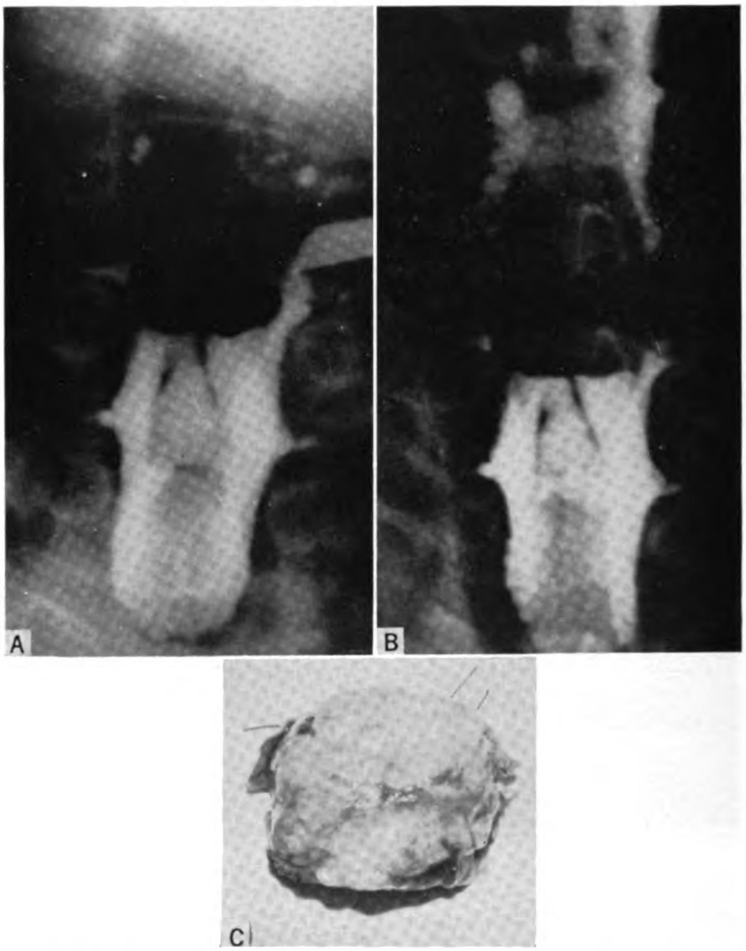

FIG. 344.—Meningioma in lower cervical spinal canal in a 34-year-old man who had a sensory level at T3 to T5, anemia and pyramidal tract signs referable to both lower extermities. A partial block was present at the level of C6, but pantopaque passed to either side. When patient was returned to the erect position the upper level of the lesion could be identified (*B*). Recovery after removal of tumor (*C*).

highly vascularized. Fine, gritty calcareous particles and sometimes larger spicules of bone or calcium are occasionally present, the calcifications occurring as a rule as small, concentrically laminated psammoma bodies. It occurs as a single lesion, but occasionally multiple meningiomas have been reported. List (1943) reported the removal of 4 meningiomas from the region of the foramen magnum and the upper cervical area. Rand (1952) reported an instance of multiple meningiomas at the sixth and the third thoracic levels. The lowermost tumor had caused a complete spinal block, and the second was found only when a catheter was pushed up beneath the dura at operation.

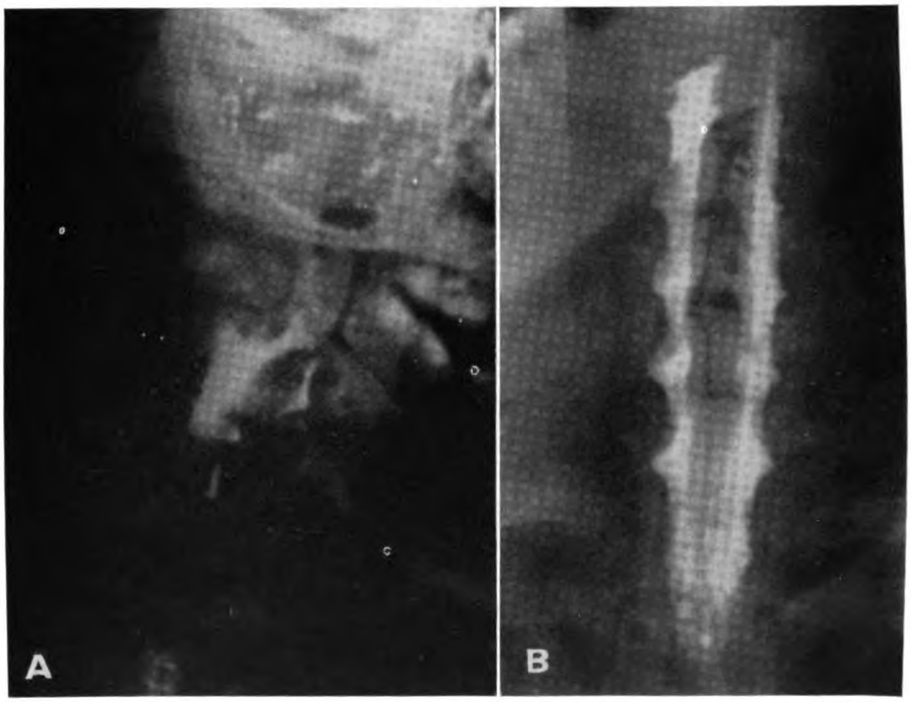

FIG. 345.—*A*, Meningioma at the level of the first, second and third cervical vertebræ. Myelography revealed an arcuate indentation at the level of the second cervical vertebra (arrow). At operation the tumor was 2 cm. long ,1.5 cm. wide and 1 cm. thick. About two-thirds lay in the spinal canal, and one-third extended into the foramen magnum. Symptoms were shoulder and arm pain, limp, and some difficulty in voiding. *B*, Another case of meningioma at the level of the second and third cervical vertebræ. Bizarre clinical picture of shoulder-arm pain and weakness of lower extremities.

High cervical meningiomas may intrude into the foramen magnum and cause increased intracranial pressure by obstructing the cerebrospinal fluid circulation. Smolik and Sachs (1954) reported 6 such cases out of 234 verified spinal cord tumors. Meningiomas in the cervical spinal canal at the first or second cervical segments are difficult to localize on the basis of the clinical picture alone. Grant (1940) mentioned that they might simulate degenerative diseases of the spinal cord. Epstein and Davidoff (1946) noted the presence of low cervical and shoulder pain as well as symptoms referable to the lower extremities in patients with high cervical meningiomas. The clinical picture encountered with meningiomas in the thoracic spinal canal is one predicated on the effect of cord pressure, and as a rule the approximate level of the lesion can be identified clinically. Tumors in the lower lumbar area may cause confusing neurologic pictures, often indistinguishable from herniated discs.

Radiologic examination of the spine in patients suspected of spinal cord tumors starts with plain roentgenograms. Dyke (1941) estimated that 25 per cent of extramedullary intradural tumors might be localized by changes in the contiguous vertebræ. Buchstein (1941) observed that plain film changes with meningiomas was considerably less than seen with perineurial fibroblastomas, only 10 per cent of the meningiomas producing visible changes as compared with 45 per cent of perineurial fibroblastomas. In our own experience small meningiomas with large neurologic deficits rarely produced bony alterations. Such manifestations were extremely useful when present, but could not be regarded as helpful in eliminating the diagnosis when absent. Psammomatous meningiomas may contain

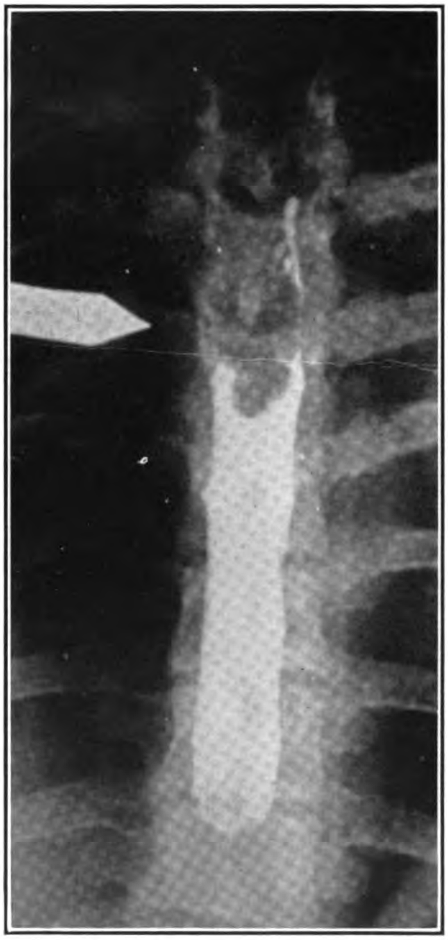

FIG. 346.—Meningioma in dorsal midportion of upper spinal canal with almost complete obstruction. Note the pantopaque flowing along the lateral aspects of the canal. The defect is centrally situated. No spreading of the lateral upward-extending streaks is present.

sufficient calcification to indicate their presence radiologically. In a series of 130 cases reported by Brown (1942), 15 per cent were either of this variety or osteoplastic in type, and 4 per cent of these were visible on plain roentgenograms. Bray (1942) suggested that body section roentgenograms might be of assistance in identifying such calcifications, which he found in about 10 per cent of meningiomas. He reported 3 cases, including 2 meningiomas, 1 of a psammomatous type and the other of the osteoplastic variety. The third case was a hemangioblastoma containing calcium. The calcifications appeared as olive-shaped opacities within the spinal canal. No associated pedicle thinning or foraminal enlargement was observed. In a case of a cervico-thoracic meningioma reported by Osgood, Arnett and Lewy (1944), the calcification measured 4 cm. in length and 2 to 5 mm. in thickness. Culver, Concannon and Koenig (1949) presented 5 cases out of a series of 15 meningiomas in which homogeneous increased density indicative of tumoral calcification was present, and was best seen on the lateral roentgenograms.

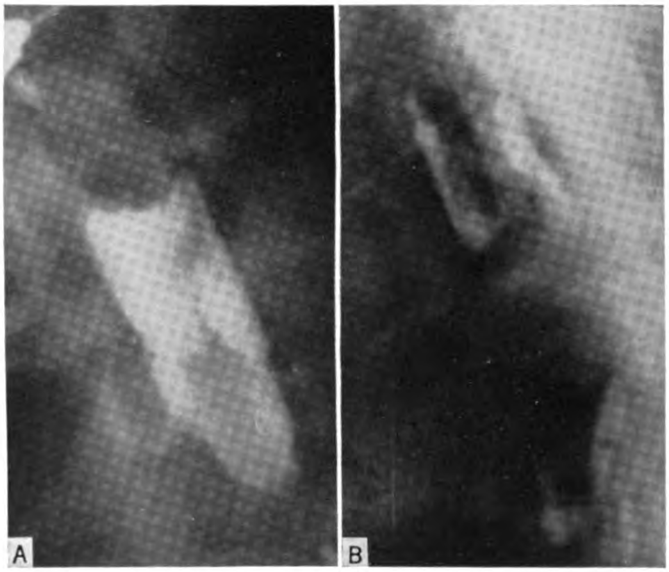

FIG. 347.—Meningioma extending for T4 to T6 in a 69-year-old woman with progressive weakness and numbness of both lower extremities for more than 18 months, ascribed to "osteoarthritis." The pantopaque column is capped at the level of T6 (*A*), but some contrast material passes the obstruction. With the patient erect, the upper aspect of the tumor is shown (*B*), and some faint calcification can be made out within the tumor. It is of interest that lumbar myelograms made at this time disclosed pronounced ridging. Inasmuch as the patient did very well after operation, the ridging was considered incidental and of no immediate significance.

Myelography is the most important single procedure in the diagnosis of intraspinal meningiomas. If a block is suspected lumbar instillation of about 1 c.cm. of pantopaque is sufficient. Tumors situated anterolaterally or posterolaterally can be identified by indentation of the column on the corresponding side. Anteroposterior localization is made on the lateral projections. Since many meningiomas are situated in the thoracic area, one must obtain an adequate flow to the upper thoracic and cervical levels. This may be rather difficult in patients with exaggerated thoracic curvatures, but this usually can be overcome by placing the patient in a lateral position. Meningiomas often produce complete or almost complete block, so that the diagnosis usually is established easily. If the uppermost extent of the lesion is to be determined in the presence of complete obstruction, cisternal instillation of pantopaque sometimes is required. This becomes quite important if multiple tumors are suspected.

If the tumor is only partially obstructive the examiner must be alert to identify small arcuate defects in the head of the pantopaque column which may be quite transient. The entire canal should be checked, up to and including the foramen magnum. The upper and lower limits of the tumor can be determined readily when partly obstructive lesions are present by observing the downward flow of pantopaque.

If an obstructive lesion is present it is better not to attempt to remove more pantopaque than can be aspirated without removing fluid as well. The possibility of a high lesion moving downward and compressing the cord must not be disregarded. For the same reason we do not perform the Queckenstedt test when gross spinal canal obstruction is suspected.

The differentiation of a meningioma from a neurinoma on myelographic evidence is not always possible. However, when a relatively sharply delimited circular or ovoid defect is present, the latter is more likely present. The extramedullary position of a mass can be identified by displacement of the negative shadow of the cord away from the tumor. If pantopaque streaks permit identification of dural attachment, a relatively elongated irregular base favors the possibility of a meningioma. Differential diagnosis between meningiomas, neurofibromas and other obstructing tumors such as myelomas or metastatic tumors is aided by the clinical picture as well as the demonstration of osseous changes. Occasionally a herniated disc must be considered, and the identification of some extradural calcification projecting toward the canal from a thoracic intervertebral disc can be helpful. In any event, myelographic diagnosis of an obstructive lesion should be made as quickly as possible after the onset of symptoms, particularly if they are sudden in onset and rapidly progressive.

The benefits of surgical removal of meningiomas are obvious. It is less well known, however, that meningiomas may recur. Three such cases were reported by Cushing and Eisenhardt (1938), recurring two years in 2 cases and nine years after removal in the third. Davidoff, Gass and Grossman (1947) reported the recurrence of a meningioma fourteen years after the removal of a lesion from the lower thoracic region. One should also recognize the possibility of other coexistent lesions. Freedman, Feiring and Davidoff (1949) reported metastases from carcinoma of the breast together with intraspinal meningiomas in three cases.

Neurofibromatosis and Perineurial Fibroblastomas.—This is a hereditary condition which occurs in both sexes about equally, and is manifest either from infancy or becomes apparent later in life. Many patients are unaware of their affliction until the appearance of characteristic tumors about the time of puberty. Neurofibromatosis is included among the neurocutaneous syndromes such as tuberous sclerosis, Sturge-Weber syndrome and von Hippel-Lindau disease. It is recognized readily in its completely developed form by the presence of multiple tumors along the peripheral nerves, skin pigmentation and a variety of concomitant congenital malformations, among which are cerebral meningoceles, congenital glaucoma, spina bifida and mental defects (Uhlmann and Grossman, 1940). A great variety of skeletal changes appear, including abnormalities in growth and

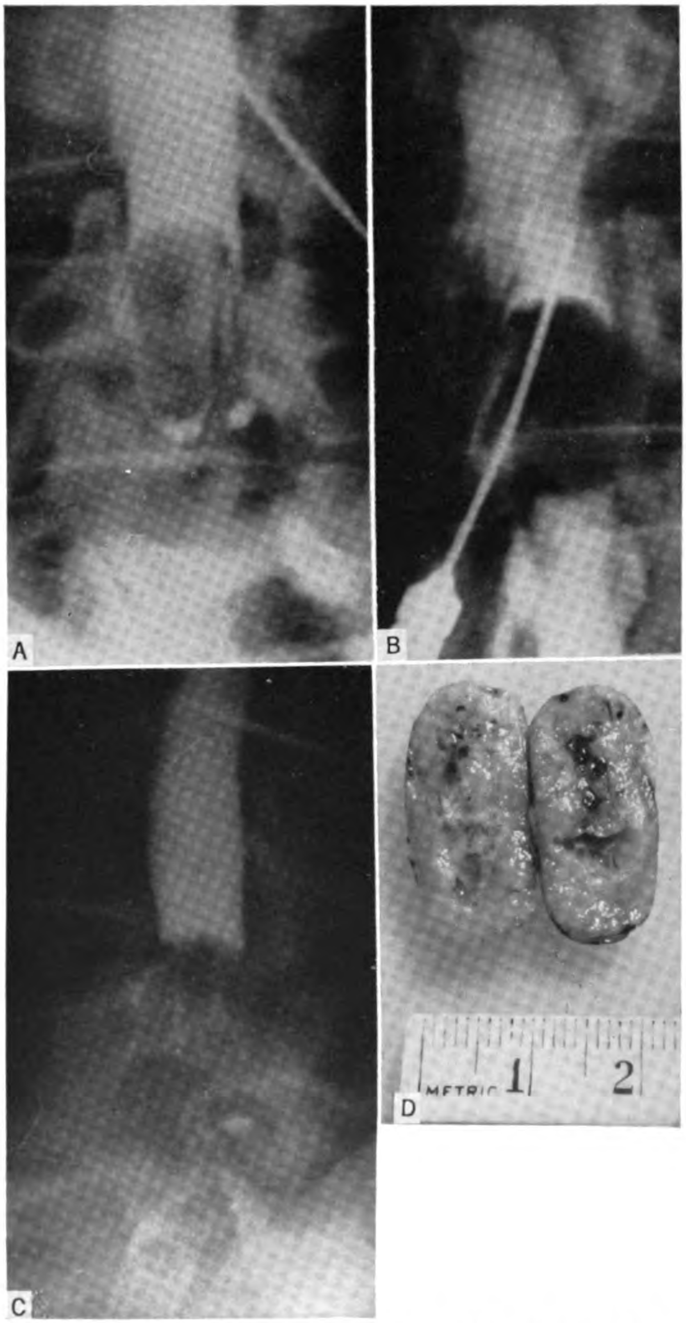

FIG. 348.—Intradural neurofibroma at the level of the fourth lumbar vertebra in a 58-year-old woman considered "neurotic" for several years. Bizarre reflex changes and intermittent pain initiated the myelographic examination. A partial block was found at the level of the superior aspect of the fourth lumbar vertebra (*A*), and a cap-like deformity was demonstrated at its upper and lower levels of the lesion (*B* and *C*). The tumor (*D*) was found attached by several filaments to one of the cauda equina nerve roots.

irregular lytic defects in the appendicular and axial skeleton. Noteworthy among these are defects in the orbital walls and the sphenoid, irregularities in growth of the tibia and fibula producing bowing, unequal lengths and pseudarthrosis of the lower limbs, soft tissue swellings and asymmetry of the face and limbs. Pronounced changes may appear in the vertebral column. Bony changes are present in about 7 per cent of cases (Friedman, 1944; Mackenzie, 1950). A variation of this condition occurs with only skin pigmentation and minor skeletal anomalies.

The appearance of neurofibromas within the spinal canal and the extension of these to the paravertebral regions is of great importance. These growths, referred to as perineurial fibroblastomas, schwannomas, neurinomas or neurillemomas, originate in the sheaths of the spinal nerve roots and occur as smooth, sometimes nodular or irregularly lobulated encapsulated growths surrounded by a firm, fibrous capsule. The tumors are the most frequent ones encountered within the spinal canal, and are usually intradural but extramedullary in location. However, sometimes they may be entirely extradural in position, and occasionally occur both intradurally and extradurally. In rare instances they may arise within the spinal cord (Walthard, 1927). They manifest themselves clinically by compression of the cord and the nerve roots. The clinical picture sometimes is complicated because of multiple intraspinal neurofibromas.

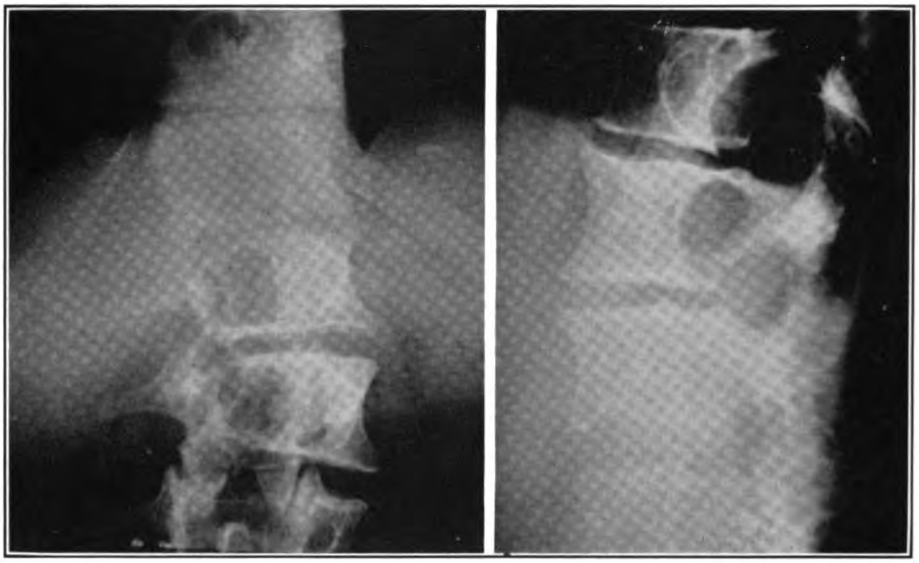

FIG. 349.—Osseous involvement in a male thirty-two years of age with generalized neurofibromatosis. Progressive weakness for several years, with paraplegia for one year before this study was done.

Perineurial fibroblastomas occur most frequently in the cervical and the thoracic spinal canal, but are not uncommon in the lumbar region. Instances of these growths within the sacral canal also have been recorded. Rasmussen, Kernohan and Adson (1940) reported that among 167 intraspinal neurofibromas, 30 were in the cervical spinal canal, 70 in the thoracic, 55 in the lumbar and 2 in the sacral spinal canal. Wolf's (1941) distribution of 67 such tumors included 25 in the cervical canal, 31 in the thoracic, 10 in the lumbar and 1 in the sacral canal. The incidence of central nervous system involvement in neurofibromatosis was reported by Holt and Wright (1948). Neurofibromatosis was present in 127 cases out of a total of 245,219 admissions. Approximately 10 per cent presented evidences of intracranial growths, a manifestation of the predilection of neurofibromatosis for localizing in the eighth cranial nerve. Their cases were studied principally from the point of view of skeletal changes, and no statistical study of intraspinal involvement was available in their report.

The full blown picture of neurofibromatosis involving the spine is characteristically one in which mild, and sometimes pronounced scoliosis or kyphoscoliosis results from malalignment of the vertebral bodies. At times a sharp angulation in the cervical spine may cause severe compressive symptoms. The lower thoracic spine is a more frequent site for such changes. In addition to alterations in the alignment of the vertebræ, the growths may produce distortion of the normal architecture of an almost cystic nature by extrinsic pressure. In some large indentations are formed in the posterior aspects of the involved bodies, usually limited to one or two vertebræ. This is produced by compression against the dorsal surfaces by large intraspinal growths. Associated with this may be atrophy of the pedicles and laminæ, and widening of the interpedicular spaces.

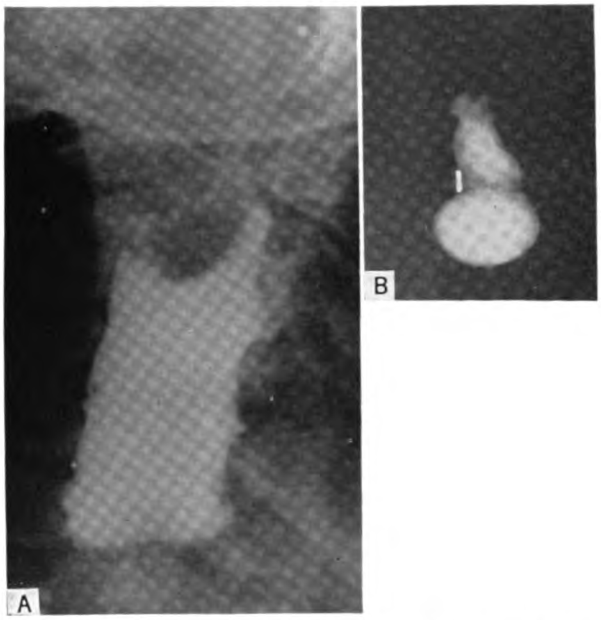

Fig. 350.—Intradural extramedullary neurofibroma in a 58-year-old woman with neck pain for one year, difficulty in walking for four months, weakness of the arms and legs and hyperactive reflexes. A smooth, cap-like indentation is seen at the level of C2 (*A*). Symptoms relieved after removal of tumor (*B*).

Of great diagnostic importance is the demonstration of widening of an intervertebral foramen, which occurs when a tumor protrudes through the foramen so that a dumbbell type of tumor results. Eden (1941) reported that in a series of 234 spinal tumors, 32 were of the dumbbell variety. Of these 25 were solitary or multiple neurofibromas, 3 were meningiomas, 2 were hemangioendotheliomas, 1 a ganglioneuroma and 1 an endothelial cell sarcoma. It is possible for the intraspinal component of such growths to be relatively small, while the extraspinal portions reach considerable size. Friedl (1946) noted that the extraspinal component may attain the size of a child's head. It is not uncommon to observe a smoothly circumscribed solid mass in the paravertebral area of the chest, usually in its upper aspect, which at first glance looks like an encapsulated growth. Associated with this may be pressure changes in the posterior aspect of the ribs. In such cases the thoracic spine must be examined to ascertain changes in the intervertebral foramina and the neural arches to avoid overlooking a dumbbell tumor. Svien (1950) noted the occurrence of 5 dumbbell tumors in 37 spinal cord neoplasms in children. One of these was a five-and-one-half-year-old child with progressive difficulty in walking, followed by paralysis of the lower extremities and bowel and bladder incompetence. A paravertebral mass was present extending from the seventh to the eleventh thoracic vertebræ. Bilateral thoracic laminectomy exposed a large encapsulated extradural tumor entering the spinal canal from the chest through the enlarged foramen between the ninth and tenth thoracic vertebræ. The intraspinal portion of the tumor was found to be a ganglioneuroma. A similar case in a three-year-old girl with calcification inside the tumor was reported by Sames (1950). Grafton and Dodge (1952) reported on 60 cases of dumbbell neurofibromas affecting the spinal canal, occurring in approximately 1000 operated cases of spinal cord tumors. They noted that spinal cord compression was the outstanding symptom in cases at the thoracic level. At the lumbar and lumbosacral levels the symptoms were those of progressive failure of the nerve roots. Cervical neurofibromas presented evidences of root failure plus spinal cord compression. In 20 of their 60 cases there were varying roentgenologic evidences of the tumor, such as increase in the size of the intervertebral foramen and erosion of the bony walls. Shortening of the pedicles and increase in the interpedicular distances, erosion of the ver-

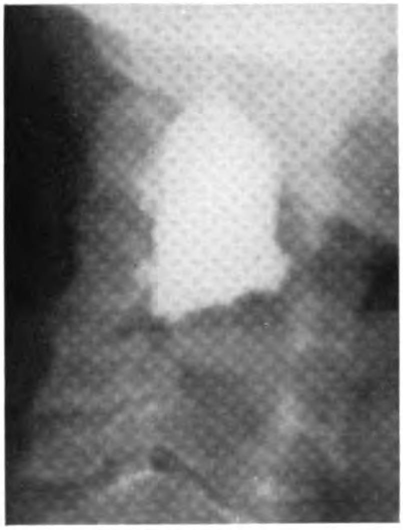

FIG. 351.—Extradural neurofibroma in a 37-year-old man with sensory changes at the level of C3 and pain in the left leg for about a year. The tumor impinged into the foramen magnum.

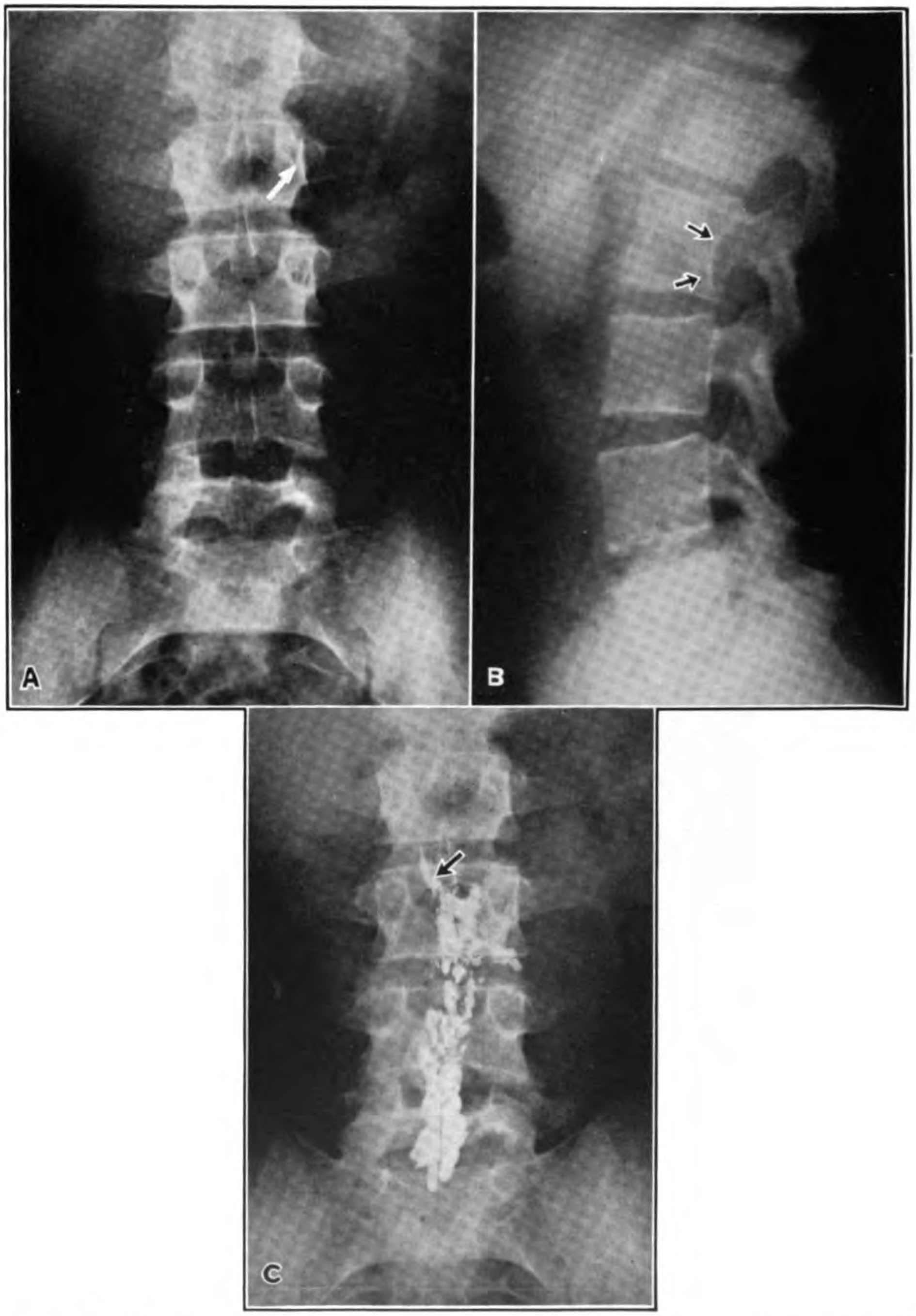

FIG. 352.—*A*, Perineurial fibroblastoma in a sixteen-year-old boy with progressive weakness and pain in both legs. The cerebrospinal fluid protein content was over 650 mgm. per cent. Anteroposterior roentgenogram shows thinning of the left pedicle of the second lumbar vertebra (arrow). *B*, Same case, lateral lumbar spine. Note the arcuate indentation of the dorsal aspect of the second lumbar vertebra (arrows). *C*, Same case. The flow of pantopaque was rather slow, and multiple irregular defects are present in the column. A curvilinear defect is present extending to the right of the midline at the upper aspect of the second lumbar vertebra (arrows). At operation a large perineurial fibroblastoma was found, and there was marked tortuosity and dilatation of the spinal cord veins below the obstruction.

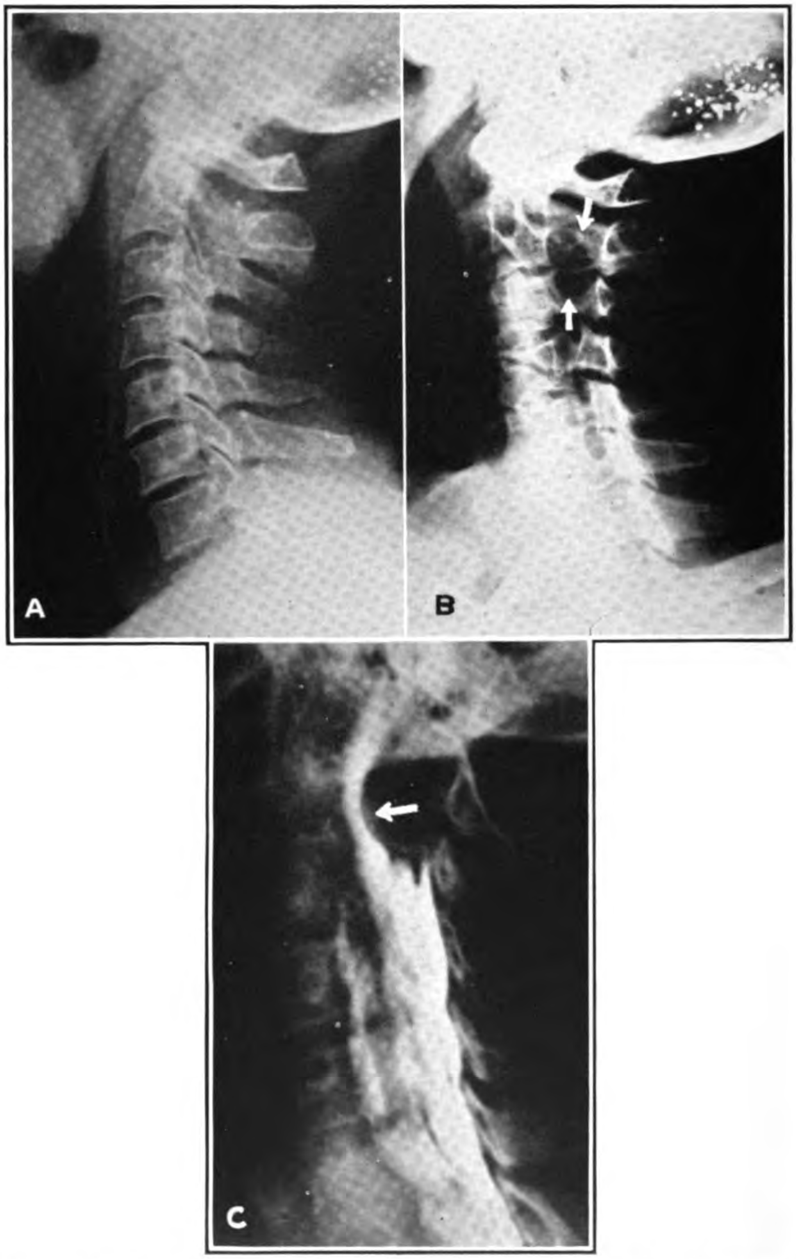

FIG. 353.—*A*, Lateral cervical spine roentgenogram in a fifty-six-year-old male with pain radiating to the occiput from the neck, and headache. No changes seen on this film. Pantopaque droplets are present in the cistoma magna. The patient's complaints had previously been attributed to this. *B*, Same case, oblique view. Note the widening of the second intervertebral foramen. (arrows). Pantopaque from a myelogram about ten years before is seen in the posterior fossa. *C*, Myelographic defect due to large perineurial fibroblastoma.

tebral bodies and laminæ, destruction of the spinous processes, erosion of vertebral portions of ribs, paraspinal masses, changes in the curvature of the spine, calcification of the tumor and hydrothorax were likewise observed occasionally. The association of neurofibromas with intrathoracic meningoceles was discussed in Chapter 2.

While neurofibromas are usually benign, neoplastic degeneration has been observed. Herrman (1950) reported 4 cases with clinical and microscopic evidences of neurofibromatosis, of which 3 developed sarcomas in their sciatic nerves and the fourth a retroperitoneal sarcoma. We had a case in which a large intrathoracic neurofibroma underwent sarcomatous degeneration, and another in which sarcomatous degeneration produced extensive destruction of the adjacent vertebræ.

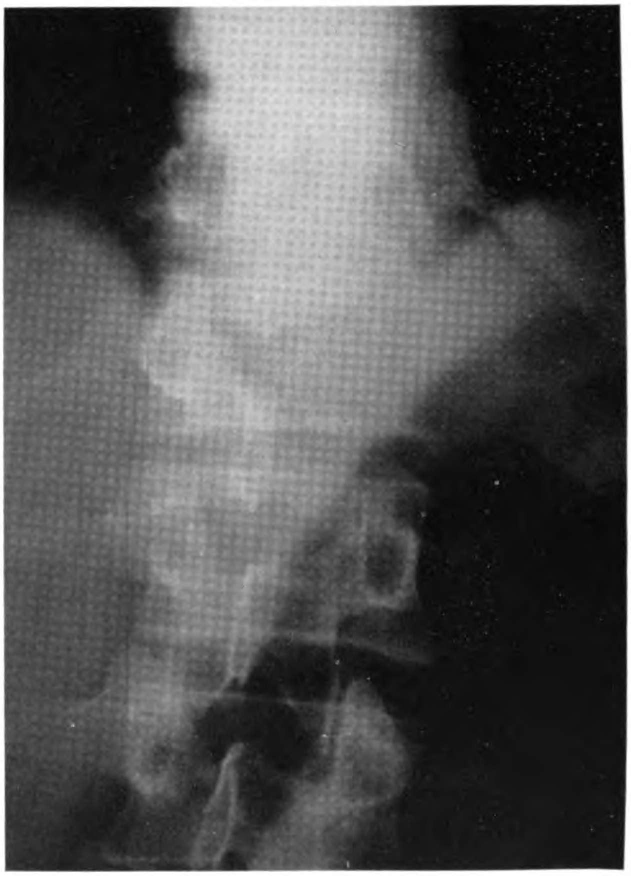

FIG. 354.—Sarcomatous degeneration of neurofibroma at the thoracolumbar junction. Patient was a thirty-five-year-old male with generalized neurofibromatosis. At operation the growth could be traced down to the spine, where it entered the spinal canal. Note the destruction of bone at the lateral aspect of the twelfth thoracic vertebra, and the adjacent rib.

Myelographic examination is required whenever clinical and roentgenologic evidence points towards intraspinal neurofibromas. In the case of an evident tumor, myelography serves to exclude multiple tumors or other masses which produce no visible or even clinical changes at that moment. The myelographic deformity may vary from a complete obstruction to the

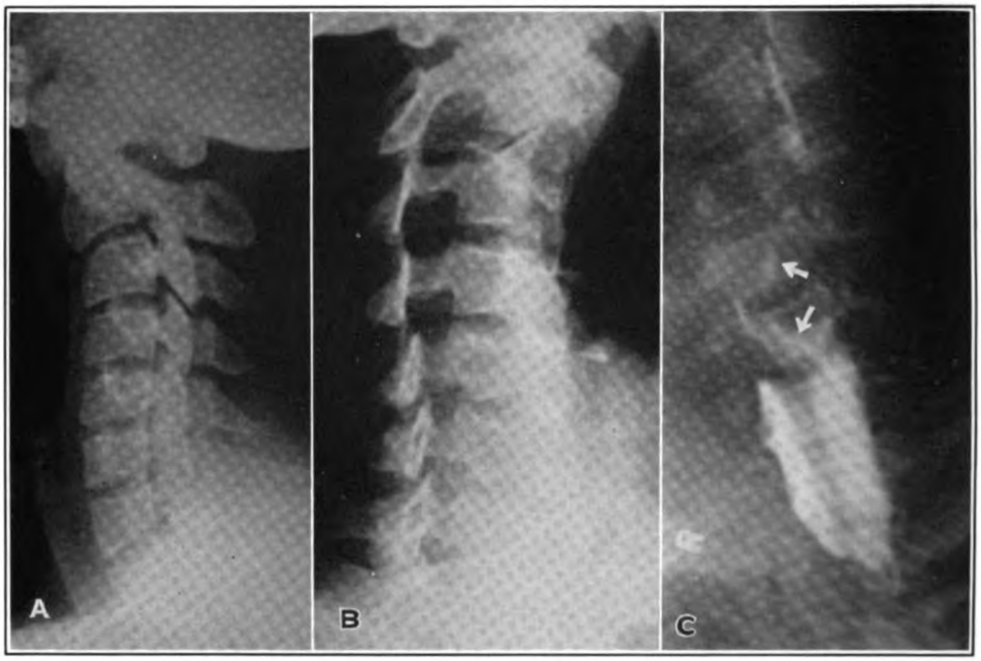

FIG. 355.—*A*, Multiple intraspinal neurofibromatosis in a twenty-nine-year-old woman. The cervical lordotic curve is straightened. *B*, Same case, oblique view. There is marked widening of the upper cervical intervertebral foramens. *C*, Same case as Figures 304 *A* and *B*, cervical myelogram. Two closely approximated curvilinear defects are seen at the lower cervical level (arrows). At operation multiple neurofibromas were found.

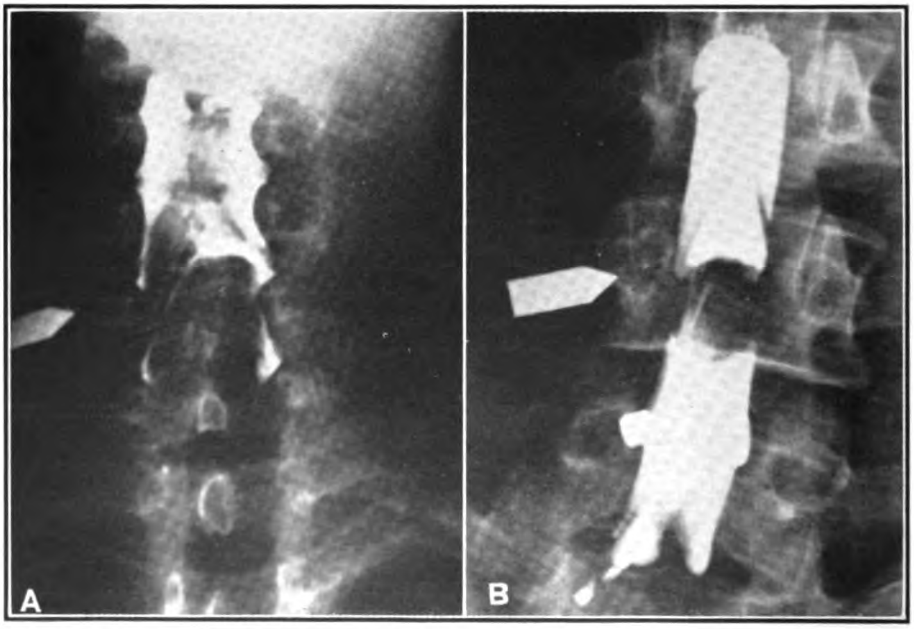

FIG. 356.—*A*, Neurofibroma in the lower cervical spinal canal. *B*, Same case. Another neurofibroma is present in the lower lumbar spinal canal.

passage of contrast medium around a sharply circumscribed circular or oval mass without obstruction. Fluoroscopic observation is essential to avoid missing a small, nonobstructive lesion which may give rise to only a transient defect before it is enveloped in the pantopaque. Wood (1949) believed that meningiomas could be differentiated myelographically from neurofibromas, because the former had broad dural attachments and slightly irregular surfaces. Neurofibromas, being encapsulated, were surrounded by the contrast medium more thoroughly than meningiomas. This is a fine differential point which in my own experience has not been entirely reliable. However, it is purely an academic distinction, inasmuch as in either event it is far more important to identify the tumor, its location and extent, than to make a histologic differentiation.

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INTRASPINAL MENINGEAL TUMOR IMPLANTS

It has long been recognized that brain tumors can spread to the spinal meninges by way of the cerebrospinal fluid circulation. Spiller (1907) described the extension of an ependymoma of the fourth ventricle to the lower thoracic region of the cord. Cairns and Russell (1931) observed spinal metastases in more than one-third of the 22 cases of cerebral glioma studied. Included in the various brain tumors were medulloblastomas, glioblastomas multiforme, astrocytomas, ependymomas and neuroepitheliomas. Half of these growths originated above and half below the tentorium. Wolf (1941) mentioned that spinal metastases from brain tumors occurred in approximately 4 per cent of all spinal cord neoplasms encountered in routine examinations of the cord at necropsy. The fact that these extensions were by means of the cerebrospinal fluid was highlighted by the report of Halpern (1942), who observed silver clips used in a posterior fossa craniotomy in the spinal canal. Three additional cases of subarachnoid and ventricular implants were reported by Tarlov and Davidoff (1946). Svien, Gates and Kernohan (1949) reported 19 cases of ependymomas of the fourth ventricle with an incidence of 31.6 per cent of spinal subarachnoid implantations. In none of these 6 patients did the history suggest the presence of implants in the canal.

While the most common site of origin for intraspinal meningeal tumor implants is from the brain, occasional cases of carcinomatosis from more distant areas occur. Meissner (1953) reported on the incidence of meningeal carcinosis in 4 cases of carcinoma of the stomach. We have seen small implants with no clinical evidence to suggest their presence in cases of leukemia, malignant lymphoma, melanoma, and breast, lung, kidney and gastric neoplasms. In a report of primary malignant melanoma of the spinal cord, King, Chambers and Garey (1952) mentioned that in their second case the cerebrospinal fluid was black, indicating the presence of free melanin. This was believed to have produced a chemical arachnoiditis which caused hydrocephalus by obstruction of the foramina of Magendie and Luschka. Spinal canal block was encountered at the level of the first lumbar vertebra, and at operation an intradural tumor was found enmeshing the cauda equina, filling the intramedullary region of the conus and penetrating the spinal cord on its anterior surface. At necropsy seeding of cells by way of the cerebrospinal fluid to the brain was found.

Unless spinal meningeal implants attain sufficient size to cause symptoms, their presence usually is detected at necropsy only. Nevertheless, there is a definitely practical point in recognizing these lesions which finds application in the use of *x*-ray therapy in cases of leukemia, medulloblastoma and other brain tumors. In children with leukemia the meninges may become seeded with tiny implants. However, leukemia and more often lymphoma are occasionally associated with tumorous masses which cause rapid spinal cord or cauda equina compression and which require prompt and adequate treatment.

Meningeal implants on the cord are more frequent on its dorsal aspect. They are usually gray, white and firm, and most of the time are tiny. Compression and distortion of the cord and thickening of the leptomeninges may be observed when the implants are sufficiently large. Another form of meningeal involvement may be tumorous thickening in which the neoplastic layer may reach a thickness of more than a millimeter.

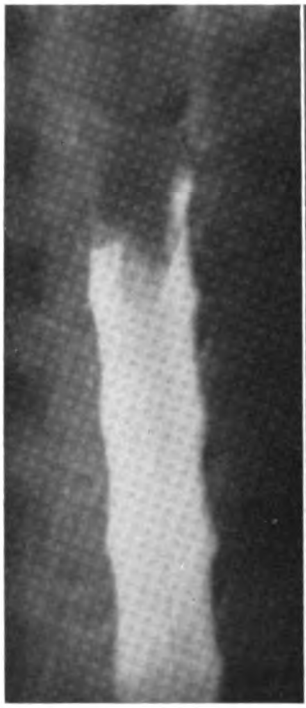

FIG. 357

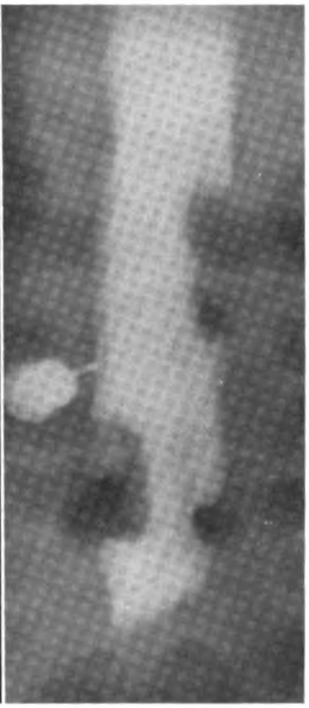

FIG. 358

FIG. 357.—Intraspinal metastases in the upper thoracic spinal canal in a 6-year-old girl operated upon two years before for a cerebellar spongioblastoma. Progressive chest pain in the lower thoracic and high epigastric areas. Manometrics normal. Relief with intrathecal methotrexate followed by *x*-ray therapy (courtesy of Dr. Leo M. Davidoff).

FIG. 358.—Intradural metastases from a cerebellar astrocystoma in a 32-year-old man operated upon five years before present examination. The clinical diagnosis had been a herniated disc. (Courtesy of Dr. Joseph A. Epstein.)

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EXTRADURAL TUMORS

These new growths may originate from the dura, its surrounding tissues, or from the nerve sheaths outside the dural reflection. They also arise from metastatic tumor implants either in the epidural space or in the adjacent bone with extension into the spinal canal. Such growths are principally from the lungs and breast, but spine metastases occur from practically anywheres in the body. While frequently associated with visible changes on roentgenograms of the spine, the probability of neoplastic extension is not excluded by a negative *x*-ray examination. It is in such instances that myelography may prove of great help in the management of the patient's complaints. Primary tumors may arise in the adjacent bony, periosteal and vascular structures, particularly sarcomas which extend into the canal

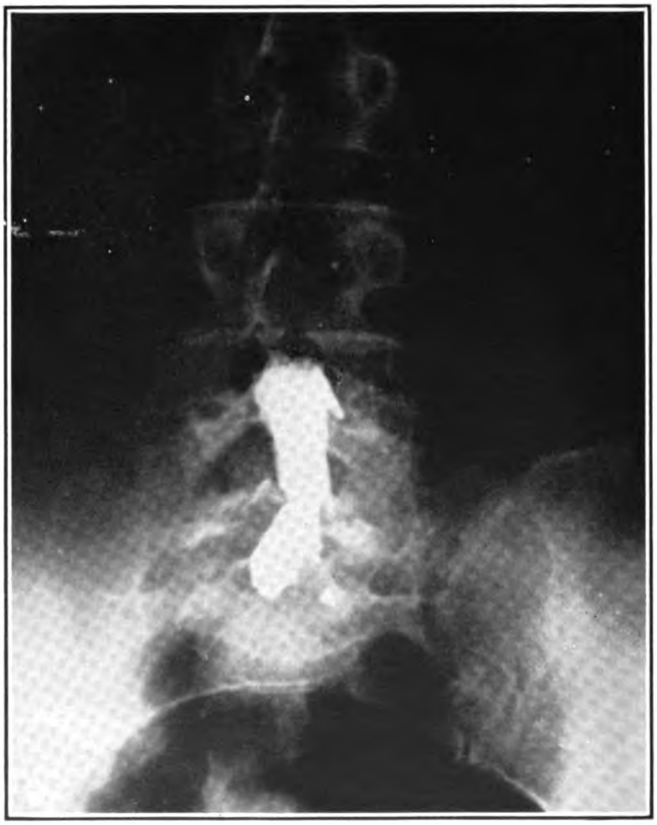

FIG. 359.—Extradural fibrosarcoma at the lumbosacral interspace in a forty-four-year-old woman who had had low-back pain for six weeks before myelography. No bony involvement noted at operation or at postmortem. Patient died in congestive heart failure due to long-standing mitral valve disease.

and produce compressive symptoms. Another tumor frequently associated with spinal cord compression is multiple myeloma. Lymphosarcoma and Hodgkin's disease may occur within the spinal canal, often without concomitant bony changes.

Inflammatory and granulomatous lesions such as epidural abscesses, tuberculomas, gummas and mycotic infections have been mentioned previously. These, too, may intrude into the epidural space. By far the most frequent condition producing epidural compression is herniation of the intervertebral disc. This, and intraspinal protrusion of osteophytes, have been discussed in the previous chapters.

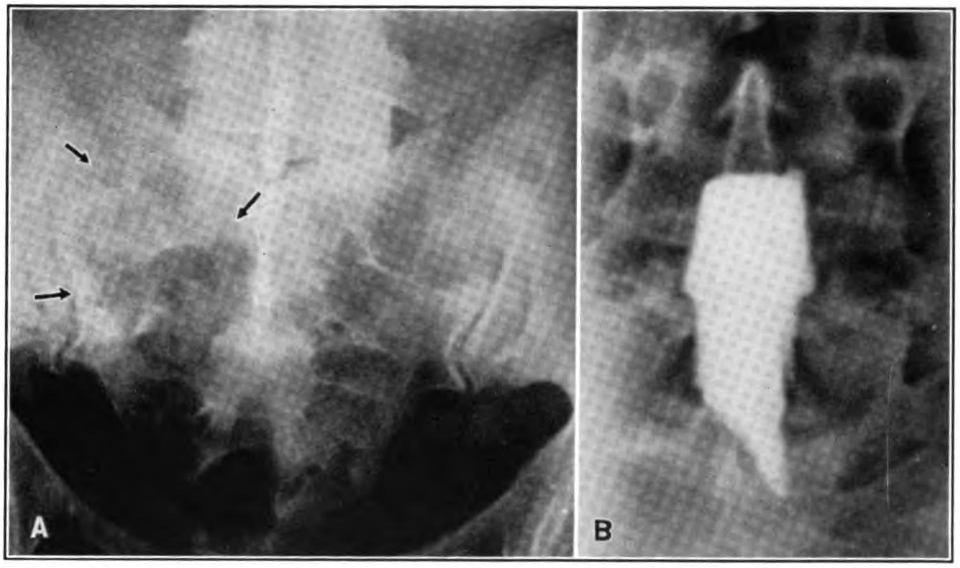

FIG. 360.—*A*, Fibrosarcoma of the upper right sacrum in a fifty-six-year-old woman with low back pain and sciatica. Mottled lytic and sclerotic areas are present adjacent to the right sacroiliac joint (arrows). *B*, Same case, myelogram showing deflection of caudal sac due to epidural extension of the tumor.

ARACHNOIDAL CYSTS

These arise as congenital diverticulæ of the dura or as arachnoidal herniations through a dural defect. They may be congenital in origin, or may follow trauma. The cysts may communicate with the subarachnoid space through a pedicle. Hyndman and Gerber (1946) reported an instance of acquired extradural cysts in the cervical region in 1 patient and in the lumbar region of another after surgery. In both small persistent fistulas in the line of dural incision permitted herniation of the arachnoid. Occasionally an arachnoidal cyst remains within the dura.

Extradural cysts are most frequent in the thoracic spinal canal, but occur occasionally in the cervical and lumbar regions as well. The walls of the cyst are composed of avascular fibrous tissue lined with epithelial cells and contain fluid similar in consistency and chemical characteristics to cerebrospinal fluid. In some of the reported cases the fluid within the cysts was somewhat gelatinous. According to DuToit and Fainsinger (1948) lumbar extradural cysts manifest themselves later than those in the thoracic region. This is ascribed to the greater capacity of the lumbar spinal canal and the ease of displacement of the cauda equina.

Congenital extradural cysts cause symptoms because of compression of the spinal cord, producing progressive paraplegia with relatively little pain. Motor weakness of the lower extremities, hyperactive deep reflexes, varying degrees of impairment of touch, pain, temperature and position sense as well as bowel and bladder disturbances may be encountered. Adams and Wegner (1947) reported intermittent compression of the spinal cord with symptoms which were subacute and progressive up to a certain point, and then regressed so that the patient was symptom free between attacks. The authors assumed that when the fluid pressure within the cyst reached

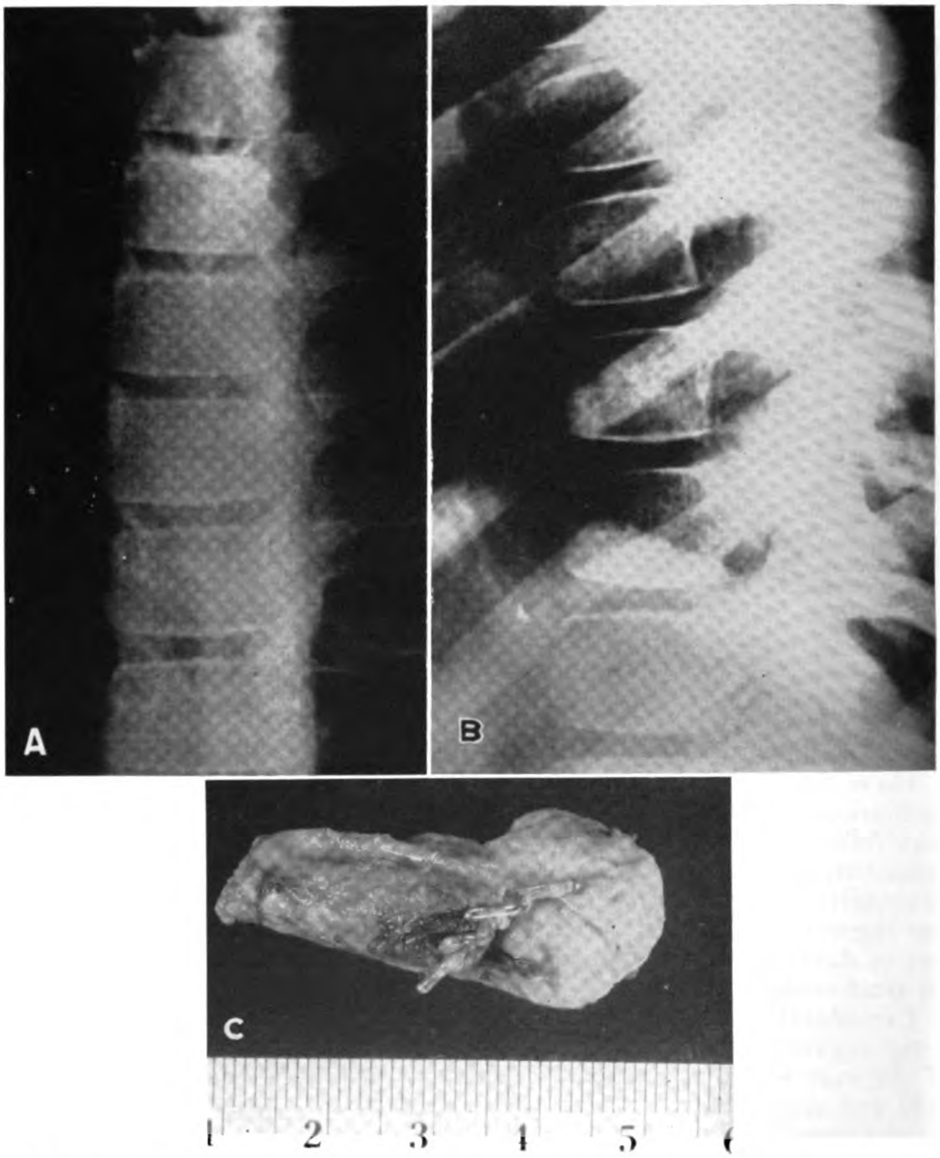

FIG. 361.—*A*, Extradural cyst in the middorsal spine of an eleven-year-old boy with progressive loss of function of both legs for five months. The interpedicular spaces are widened (arrows), with atrophy of the pedicles. *B*, Lateral dorsal spine, same case as *A*. No evidence of Scheuermann's disease is present. *C*, Operative specimen.

a certain height its walls ruptured, thereby relieving the compression. Another possibility was absorption of the cyst contents followed by reaccumulation. Swanson and Fincher (1947) reported 4 acquired extradural arachnoidal cysts complicating 1,700 laminectomies. Three of these had had previous lumbar laminectomies for herniated discs. The fourth was related to a nonpenetrating injury of the lumbar spine. Swanson and Fincher held that the predisposing cause was a dural tear in which a watertight closure could not be made. The clinical aspects of their cases suggested a recurrent intervertebral disc prolapse. Multiple needle punctures with a large spinal needle may also be a cause of extradural arachnoidal cysts. The case reported by Turner (1947) showed herniation of the arachnoid with a characteristic pedicle passing through a sharply delimited dural defect.

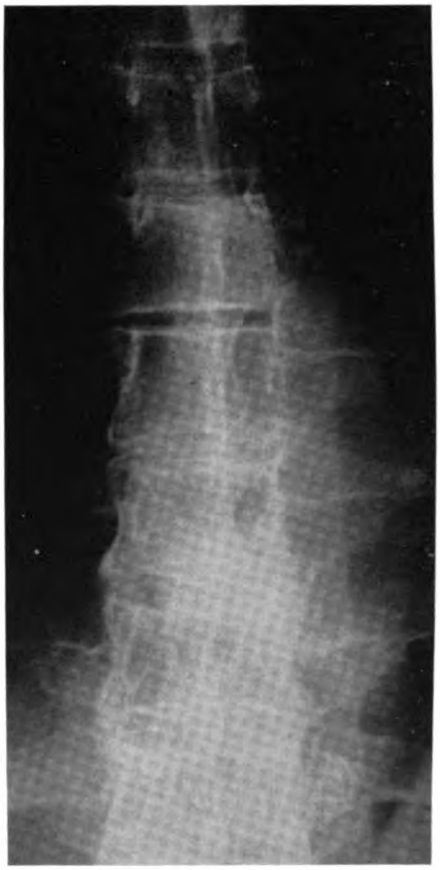

FIG. 362.—Extradural cyst in midthoracic spine of a sixty-one-year-old man. At operation the cyst communicated with the subarachnoid space. The pedicles are thinned and the interpedicular spaces widened in a fusiform manner at the levels of the sixth to tenth thoracic vertebræ.

The plain film changes seen with arachnoidal cysts reflect the gradual increase in the size of the fluid-containing sac and its pressure against the adjacent bone. The earliest change is slight loss of bone in the inner aspects of the adjacent pedicles. This affects a single vertebra when a small cyst exists, while with larger sacs the pedicles of several segments become thinned and widened. Usually the pedicles at the upper and lower limits of the lesion are less altered than those in the middle where the bulk of the cyst is largest. As the cyst increases in size indentations appear against the posterior aspects of the vertebral bodies resulting in scalloped vertebræ. The neural arches likewise may become thinned. Usually these changes can be differentiated from ependymomas easily when they appear in the cervical or thoracic spinal canal on the basis of localization alone. In the lumbar region the changes sometimes are difficult to distinguish from a bulky tumor.

Myelography is helpful in differential diagnosis, and provides an almost pathognomonic sign when some of the contrast material, be it pantopaque or air, enters the cyst. In some instances the neck of the cyst permits such communication, but in others this does not occur. It is sometimes helpful to repeat the films after a few hours to see if air or pantopaque enters the sac. When pantopaque envelopes the lesion it produces a partial block at each level where the cyst impinges against bone, and some a complete block is produced. A localized small cyst, such as may be seen with a circumscribed area of arachnoiditis, may produce only a slight lateral filling defect simulating a herniated disc. Hart (1958) suggested that if operation did not disclose a discal lesion to explain the myelographic defect it might be necessary to open the dura.

Plain film roentgenograms in children and adolescents may show concomitant changes of Scheuermann's disease (Cloward and Bucy, 1937). Adelstein (1941) observed that the association of Scheuermann's disease with progressive paraplegia indicated a spinal extradural cyst. None of the cases of Scheuermann's disease seen by us had extradural cysts.

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PERINEURIAL CYSTS AND MENINGEAL DIVERTICULÆ

Tarlov (1948) described perineurial cysts as originating from the posterior sacral nerve roots, as a rule 1 cm. or more from the origin of the nerve root from the thecal sac. He considered them a cause for sciatica. Inasmuch as they did not communicate with the subarachnoid space, they produced no changes either on plain films or on myelograms. Tarlov reported that he had observed such cysts in 5 out of a total of 30 cases in which the spinal cord was examined at necropsy.

Schreiber and Haddad (1951) mentioned that 1 of each hundred patients operated upon with a diagnosis of lower lumbar disc rupture had lumbar or sacral cysts filled with cerebrospinal fluid. They reported 4 which arose as fluid-filled swellings within the nerve, or emerged from the dura as meningoceles compressing the cauda equina. Plain films showed some thinning of the overlying bone. Strully and Heiser (1954) also differentiated lumbar and sacral cysts of meningeal origin from those described by Tarlov. In 4 verified cases microscopic examination showed the cysts to be lined with arachnoid rather than composed partially of axis cylinders as described by Tarlov. Jacobs, Smith and van Horn (1954) published myelograms showing pantopaque trapped in multiple pockets at the first and second sacral nerve roots. These were about a centimeter in diameter and about the same distance from the spinal canal. At operation small cysts containing clear cerebrospinal fluid were found.

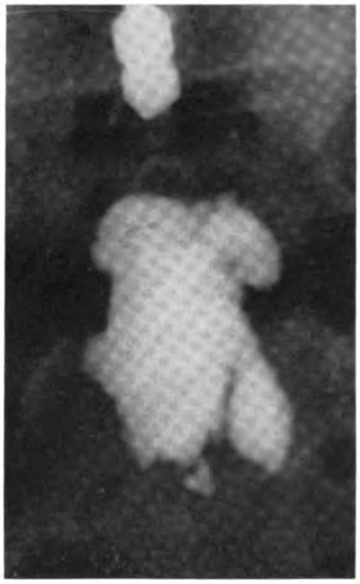

FIG. 363.—Multiple large arachnoidal diverticulæ, asymptomatic, in a patient suspected of a cervical spinal cord tumor. Myelographic examination otherwise normal.

Taheri, Riemenschneider and Ecker (1952) reported a case of multiple perineurial cysts in a thirty-eight-year-old woman with intermittent low-back pain in whom a myelogram disclosed pantopaque entering the terminal portion of the sac. The pantopaque was entrapped inasmuch as it did not return to the main column when the patient was tilted. This cyst was removed after the sacrum was unroofed, and the patient was relieved of pain.

In our own experience we have observed 50 cases in which these grape-like projections were noted either at the distal end of the caudal sac or along the third to fifth nerve roots at points of from 1 to about 2.5 cm. from the main column. In many the pantopaque returned to the subarachnoid space with changes in position. In a few the pantopaque remained within the small sacs. There was great hesitation on the part of the neurological surgeons to intervene on the basis of this change alone, but it is conceivable how they may produce pressure against the nerve roots and thus cause symptoms. In a few instances the sacs did not fill during myelography, but films made a few days later showed these pantopaque filled protrusions when some of the contrast medium remained in the spinal canal.

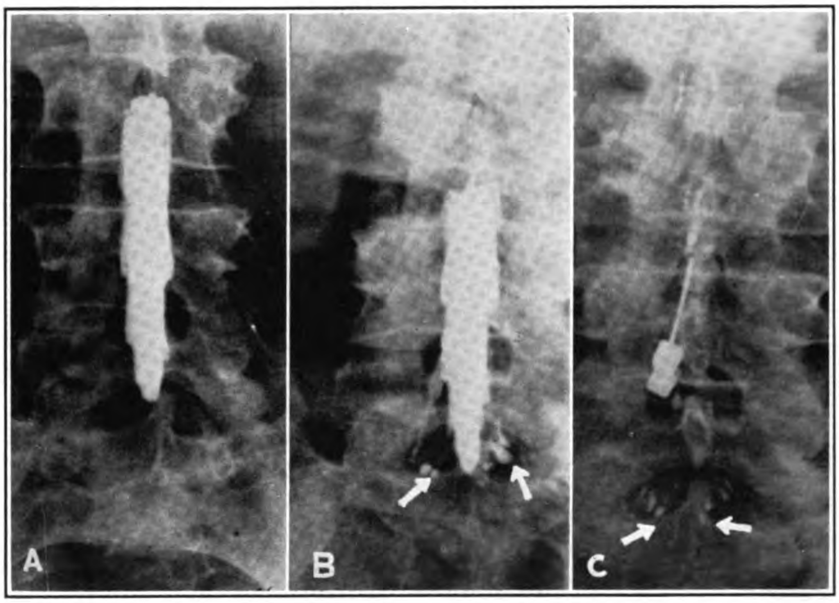

FIG. 364.—*A*, Normal myelogram with high and narrow caudal sac. *B*, Same case as *A*, about five minutes later. Pantopaque has entered several grape-like arachnoidal spaces (arrows). *C*, Same case after removal of pantopaque. Some contrast medium remains in the arachnoidal diverticulæ (arrows).

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INTRASPINAL VASCULAR MALFORMATIONS

Vascular malformations within the spinal canal are either venous, arterial or arteriovenous in origin. They involve principally the leptomeninges, and vary in extent from single convoluted dilated vessels to huge intertwined varicosities. Occasionally a bruit can be heard over the involved area. Vascular neoplasms such as capillary hemangiomas, hemangioendotheliomas, hemangioblastomas and hemangiomas associated with vertebral angiomas also project into the spinal canal, but cannot be considered as true vascular malformations. Dilatation of the venous structures may result from intraspinal lesions causing obstruction to the return flow of blood. The presence of such venous dilatations resulting in varices was exemplified by a patient of Hare and Everts (1937) who developed calcification within the spinal canal after an injury. At operation twenty-five years later varices were found above the calcified area. The presence of dilated veins above a large herniated disc or intraspinal tumor is well known to the neurological surgeon. The effects of vascular dilatations within the spinal canal depends on their extent. Minute dilatations frequently exist without producing symptoms (Kaydi, 1929).

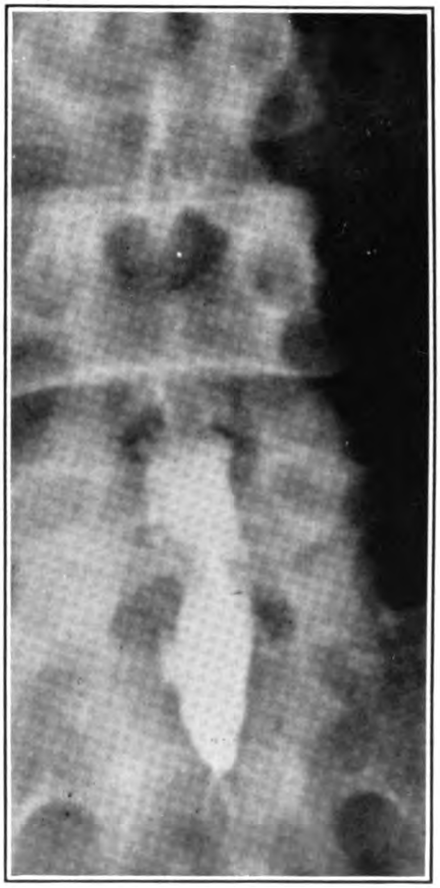

FIG. 365.—Myelographic defect on the left side at the lumbosacral interspace due to local epidural varix. The clinical picture was that of a herniated disc in a fifty-two-year-old man.

Local dilatations of epidural veins, particularly in the lumbosacral region, may compress adjacent nerve roots and produce a clinical and radiological picture indistinguishable from herniated discs (Cohen, 1941). Epstein (1947) reported 3 such cases, and others have been mentioned in the course of reports on intervertebral disc herniations (Johnson, 1940; Maltby and Pendergrass, 1946).

The association of subarachnoid angiomas with subcutaneous hemangiomas of a corresponding metamere was originally described by Berenbruch (1895), who demonstrated an actual vascular connection through the intervertebral foramen between an angioma of the cervicothoracic portion of the cord and multiple angiolipomas of the skin of the upper part of the trunk. Turner and Kernohan (1941) mentioned that instances of vascular tumors of the cord associated with nevi of the corresponding cutaneous metamere was reported only four times prior to 1941. In their study of 46 cases of vascular tumors of the spinal cord there were 18 angiomas, in none of which was there a vascular or pigmented nevus. Cross (1948) described a case of cutaneous and intraspinal angioma in the cervical region of a nineteen-year-old male which he believed to be the sixth of this condition. Trupp and Sachs (1948) also had a patient with a large skin telangiectasis who suddenly became paraplegic at a level corresponding with the skin lesion. It is probable that this group falls into the classification of Lindau's disease,

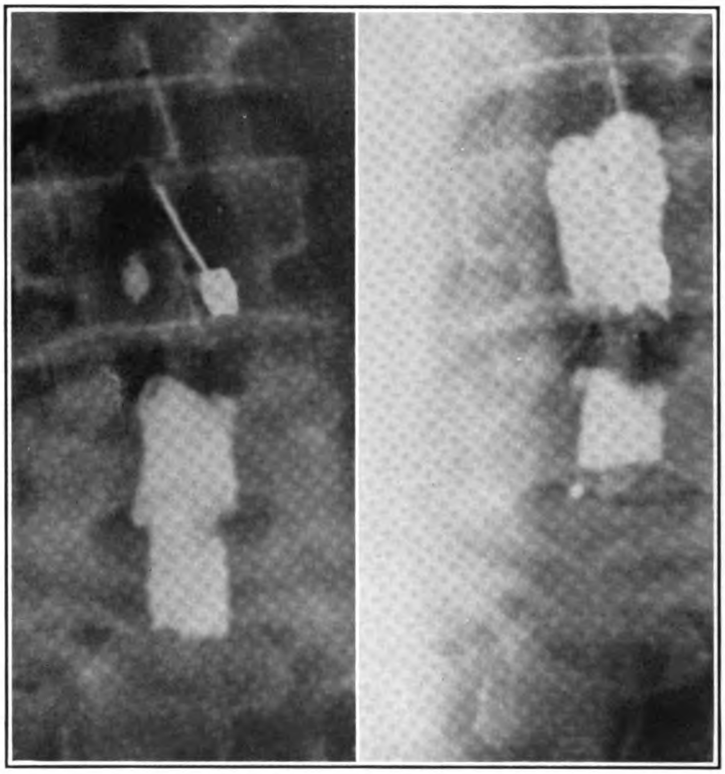

FIG. 366.—Transverse myelographic defect at the fourth lumbar interspace due to large epidural varicose veins. The clinical picture was that of a herniated disc. No disc found at operation.

consisting of familial hemangiomatous cysts, tumors of the central nervous system, associated angiomatosis of the retina and cysts of other parenchymatous organs. As brought out by Craig and Horrax (1949), these angiomatous lesions may extend the entire length of the cord. A photograph of such a specimen was published by Davidoff and Epstein (1955).

Vascular malformations within the spinal canal may produce obstruction and cord compression simulating a spinal cord tumor or a herniated disc. Elsberg (1925) cautioned against accepting varices as a sole cause for such a syndrome without searching for a tumor or other obstruction at a higher level which might cause venous obstruction and dilatation.

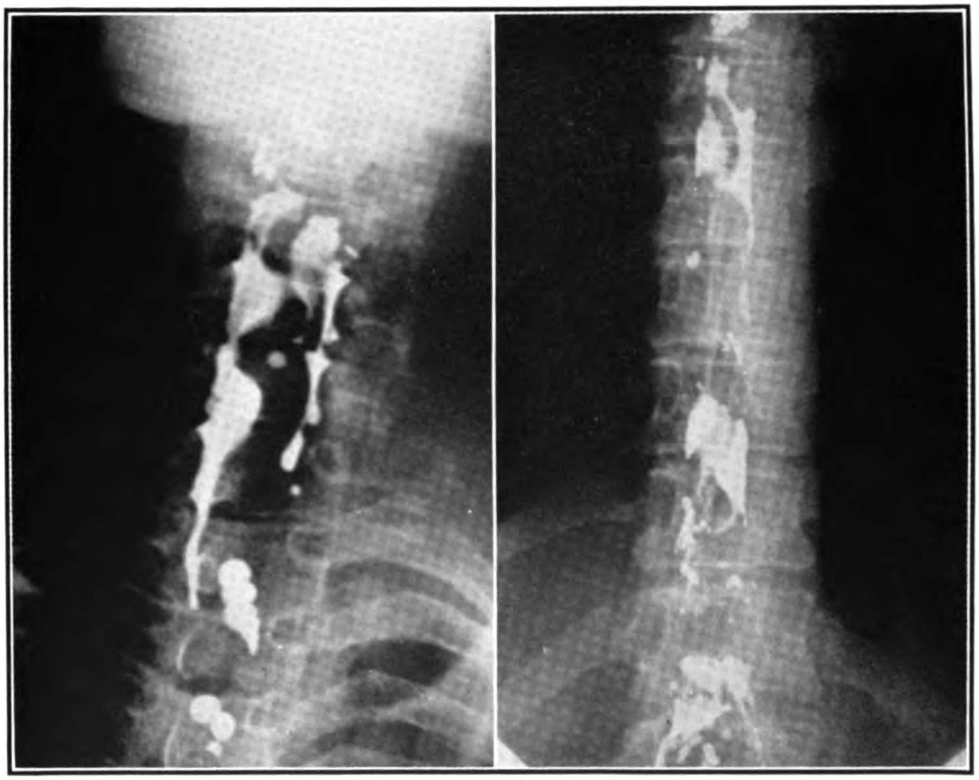

Fig. 367.—Multiple channels due to extensive arachnoidal varicosities. (From Epstein, B. S. and Davidoff, L. M., Am. J. Roentgenol., **55**, 413, 1946, courtesy of Charles C Thomas, Publishers.)

When an intraspinal angioma bleeds, there may be intense sudden pain in the back with radicular extension along the involved nerve roots. Syntoms may be intermittent and residual discomfort follows scar formation with nerve root pressure. Bassett, Peet and Holt (1949) stressed that at this stage the objective signs may be bizarre and segmentally incompatible with a single lesion because of the multiple foci of irritation. Consequently, the clinical picture may simulate multiple sclerosis, poliomyelitis or other inflammatory or degenerative diseases. Epstein and Davidoff (1943) pointed out that these symptoms of exacerbation and remission may indicate variations in the degree of vascular dilatation at any particular moment.

Small intramedullary vascular malformations may rupture and produce intramedullary bleeding, with sudden onset of pain and rapid development of long tract signs. The bleeding may be confined to the cord, resulting in hematomyelia. If no blood enters the subarachnoid space spinal puncture produces only clear fluid. Myelography is required for determination of the site of the lesion, which can be demonstrated anatomically if obstruction is produced or if a localized swelling in the cord can be identified. Under such circumstances the lesion may not be diagnosable until revealed at operation.

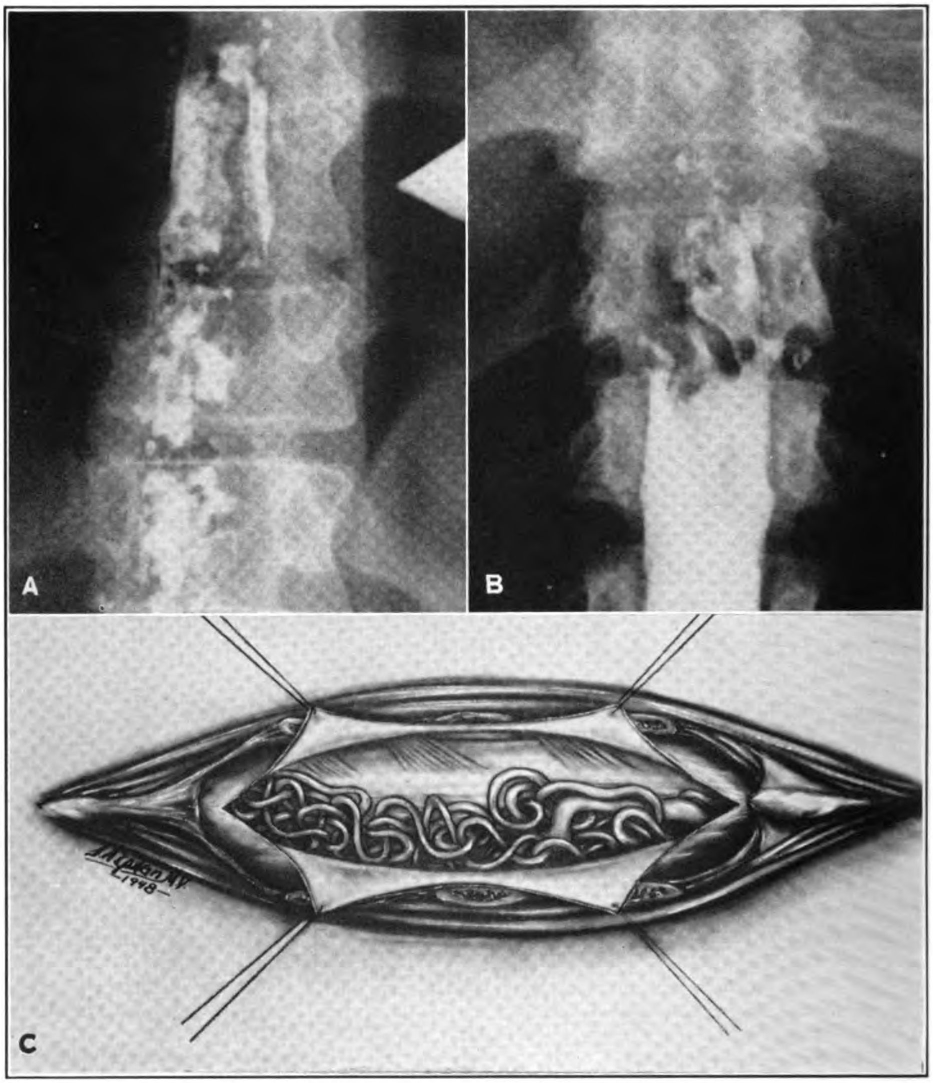

FIG. 368.—*A* and *B*, Myelographic defects due to arachnoidal arterial anomalies. *C*, Same case operative drawing (Epstein, J A , Beller, A J , and Cohen I , courtesy of J Neurosurg., **6**, 45, 1949)

X-ray examination of the spine in cases of intraspinal vascular abnormalities are usually normal. However, if a localized mass of sufficient size exists, there is evidence of local bone pressure atrophy. Actual widening of the spinal canal in the course of an intradural hemangioma and aneurysm with calcification was described by Walker (1944) in his discussion of congenital anomalies of the spinal cord. The interpedicular spaces in this patient were widened, but no erosion was present, and it was assumed that the altered configuration of the spine was part of the congenital anomaly.

In the presence of spinal cord varices, myelography is of great diagnostic importance. Pressure of the dilated vascular channels in the pantopaque column results in sinuous, radiolucent channels which are quite characteristic. Their appearance may vary from moment to moment depending on the degree of filling at the time. If only a single focal lesion is present, the myelographic picture may be only a single defect in the pantopaque column without any distinguishing characteristics.

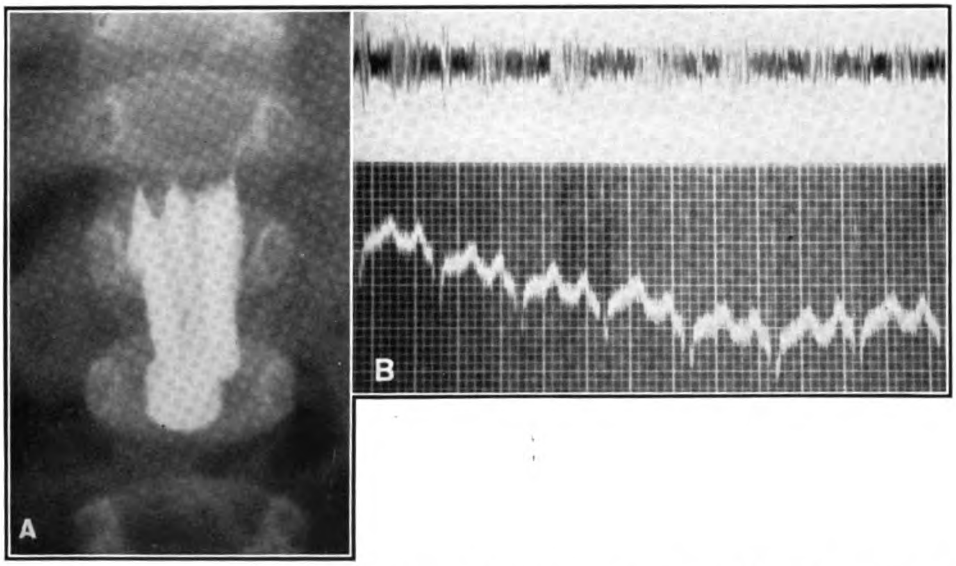

Fig. 369.—Arteriovenous malformation in an 8-year-old girl with symptoms of a spinal cord tumor at the level of the twelfth thoracic vertebra. Myelography (*A*) disclosed a complete block at this level. On auscultation a murmur could be heard at this site, confirmed by phonocardiogram (*B*) and at operation. (Courtesy of Dr. Joseph A. Epstein, North Shore Hospital, Manhasset, N.Y.

It is not possible to differentiate arterial, venous or arteriovenous malformations radiologically (Epstein, Beller and Cohen, 1949). It has been mentioned that there may be difficulty in differential diagnosis between vascular anomalies and adhesive arachnoiditis. In our experience, and from reviewing the published roentgenograms of others, the sinuous, smooth-walled channel-like structures of vascular dilatations are rarely simulated by the more diffuse changes of arachnoiditis. Bassett, Peet and Holt (1949) pointed out that adhesive arachnoiditis might be superimposed on angiomatous malformations as a result of organized hemorrhage.

Matthews (1959) mentioned the diagnostic importance of an audible bruit over intradural spinal angiomas in two cases. A similar observation is documented in Figure 369.

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SPINAL EPIDURAL HEMORRHAGE

This usually occurs after trauma or strain, with a rapid or gradual onset of spinalcord compression. It has also been associated with anticoagulant therapy (Alderman, 1956). As a rule the hematoma is dorsal in position (Lougheed and Hoffman, 1960). Yaskin and Groff (1941) reported a case following slight trauma, with slowly progressive symptoms due to epidural hemorrhage in the upper thoracic region. The plain films of the spine were normal, but cerebrospinal fluid manometric measurements indicated a partial block. At operation a hematoma 4 cm. long and from 1.5 to 2 cm. thick was removed. VerBrugghen (1946) mentioned 8 cases from the literature, and described another in a seventy-five-year-old man following indirect trauma. In this case the short progressive history with gradual onset of paraplegia and the absence of a bony lesion suggested the presence of an extradural hemorrhage. Svien, Adson and Dodge (1950) also reported a case following trauma in which the plain films were normal, but myelograms disclosed a block at the superior margin of the third lumbar vertebra which was diagnosed as a herniated disc. At operation a hard fibrous mass compressing the dural sac was encountered. When this was opened old blood exuded, and examination of the excised sac showed an old organized hematoma. This patient made an uneventful recovery. Schultz, Johnson, Brown and Mosberg (1953) collected 12 cases and reported 4 of their own. They also noted the association with mild trauma, and corroborated the clinical similarity with lumbar disc lesions.

The neurologic findings depend upon the extent of the hemorrhage. When massive bleeding occurs motor and sensory impairment is rapid, requiring immediate operation. The lesions may be extensive. One of the cases reported by Schultz, Johnson, Brown and Mosberg (1953) presented a sausage-shaped mass extending from the ninth thoracic to the first lumbar vertebræ, involving the dorsal and right lateral aspects of the neural sac. In their review of the literature they found 3 cases confined to the cervical area, 2 of which were limited to the cervical levels. The third extended from the third cervical to the lumbar level. There were 4 lesions in the upper dorsal area, and 2 of the lower dorsal area, 1 extending to the first lumbar level. In 3 cases the lesions were confined to the lumbar region.

Plain film roentgenograms of the spine are rarely helpful unless a vertebral injury provides a lead to the diagnosis. Myelography is most important. Occasionally the spinal tap provides a lead to the diagnosis, particularly when one is sure that the needle is properly placed and the tap is dry or productive of bloodstained fluid. If this occurs it may become necessary to introduce the pantopaque into the cisterna magna. Usually the myelographic picture is one of partial or complete block, but no specific pattern is identifiable. In some instances, particularly when complete obstruction is present, the configuration suggests the possibility of an intramedullary tumor by the spreading of the head of the column. There may be considerable difficulty in diagnosis when no block is present, as occurred in 2 out of the 24 cases reviewed by Lowrey (1959).

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SPONTANEOUS SUBARACHNOID INTRASPINAL HEMORRHAGE

Spontaneous subarachnoid hemorrhage with intraspinal tumors may give rise to severe pain in the lumbar region extending down both lower extremities, accompanied by violent headache. Abbot (1939) reported the case of a sixteen-year-old boy who experienced sharp pain in the lumbar region and both lower extremities after attempting a high jump. Severe headache followed, with weakness and flexor spasm of the legs, nuchal rigidity, positive Kernig and Brudzinski signs, but no paralysis. Examination of the cerespinal fluid showed blood but no block. The patient recovered rapidly, but had over 25 similar attacks in the next fourteen months. Myelography finally disclosed a block at the level of the first lumbar vertebra. A large encapsulated ependymoma attached to the filum terminale was found at operation, with an unusual amount of hemorrhage.

Fincher (1951) called attention to the similarity of spontaneous subarachnoid hemorrhage in intradural tumors with subarachnoid bleeding from other causes. He mentioned that if such a picture were associated with radicular pain of the lower limbs, a lumbar tumor should be considered. Early myelography was recommended for detecting such a lesion before compressive symptoms or destructive spinal cord and nerve root changes appeared. He reported 5 such cases, including 3 ependymomas, 1 neurofibroma and 1 neuroglioma. In 1 of his cases myelograms showed a complete block although repeated manometric studies had not indicated this condition.

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Chapter

9

Diseases of the Hemopoietic, Collagen and Reticulo-endothelial Systems

THE LEUKEMIAS

It is generally accepted that the leukemias are essentially neoplastic diseases which originate in the blood-forming organs, from which abnormal white blood cells of varying maturity and quantity are discharged into the peripheral circulation. A close relationship between the leukemias and lymphomas has been indicated by hematologic and pathologic investigations. Willis (1952) regarded the leukemias as circulating tumors, and felt that heterotopic infiltrations and tumors were true metastatic growths arising from the blood-borne malignant leukocytes. From the point of view of the roentgenologist, no sharp differentiation can be made in the osseous radiologic manifestations of the various types of leukemia. In children, the acute leukemias are often accompanied by more marked osseous manifestations. A summary of the present-day clinical status of the lymphomas and leukemias was made recently by Rosenthal (1954). Lymphomas of the vertebral column and spinal cord were considered in Chapter 5.

The osseous manifestations of the leukemias appear most frequently in the appendicular skeleton of children. Caffey (1950) regarded these as due most likely to malignant proliferation of reticulum cells originally native to the bone marrow, rather than to malignant proliferation of reticulum cells transported from lymphatic structures such as the spleen or lymph nodes. However, Willis (1952) concluded that the leukemias are malignant neoplasms with permanent circulating metastases from a tumor originating in the blood-forming tissues. Hence the heterotopic infiltrations and tumors are true metastatic growths arising from blood-borne malignant leukocytes. In early stages the marrow cavity may be packed with leukemic cells, and pain may follow such increased pressure. These changes usually are of a lytic nature. Occasionally there may be evidence of osteosclerosis, which is due to an increase in the density of the spongiosa rather than cortical thickening. At times subperiosteal new bone formation occurs particularly when there has been perforation of the inner aspect of the cortex by leukemic spongiosal infiltrations with subsequent elevation of the periosteum. One occasionally finds periosteal thickening secondary to subperiosteal hemorrhage and elevation of the periosteum without underlying cortical perforation. In the long bones, the most characteristic change is a zone of rarefaction immediately proximal to the metaphyses. This change was noted in 70 per cent of 43 cases of childhood leukemia reported by Bary and Vogt (1935), who could find no clear explanation for the production of this zone. They postulated that infiltration and erosion of bone by masses of leukemic cells probably played an important part in the production of this rarefied metaphyseal zone. Silverman (1948), in a report of 103 infants and children with leukemia, mentioned that this was the most common change observed.

No mention was made of spinal leukemic changes in the cases reported by Silverman (1948). In a report of 168 cases, Craver and Copeland (1935) observed that there were 6 with demonstrable changes in the femurs, sternum, humerus, pelvis, skull, metacarpals and ulna and vertebræ, in that

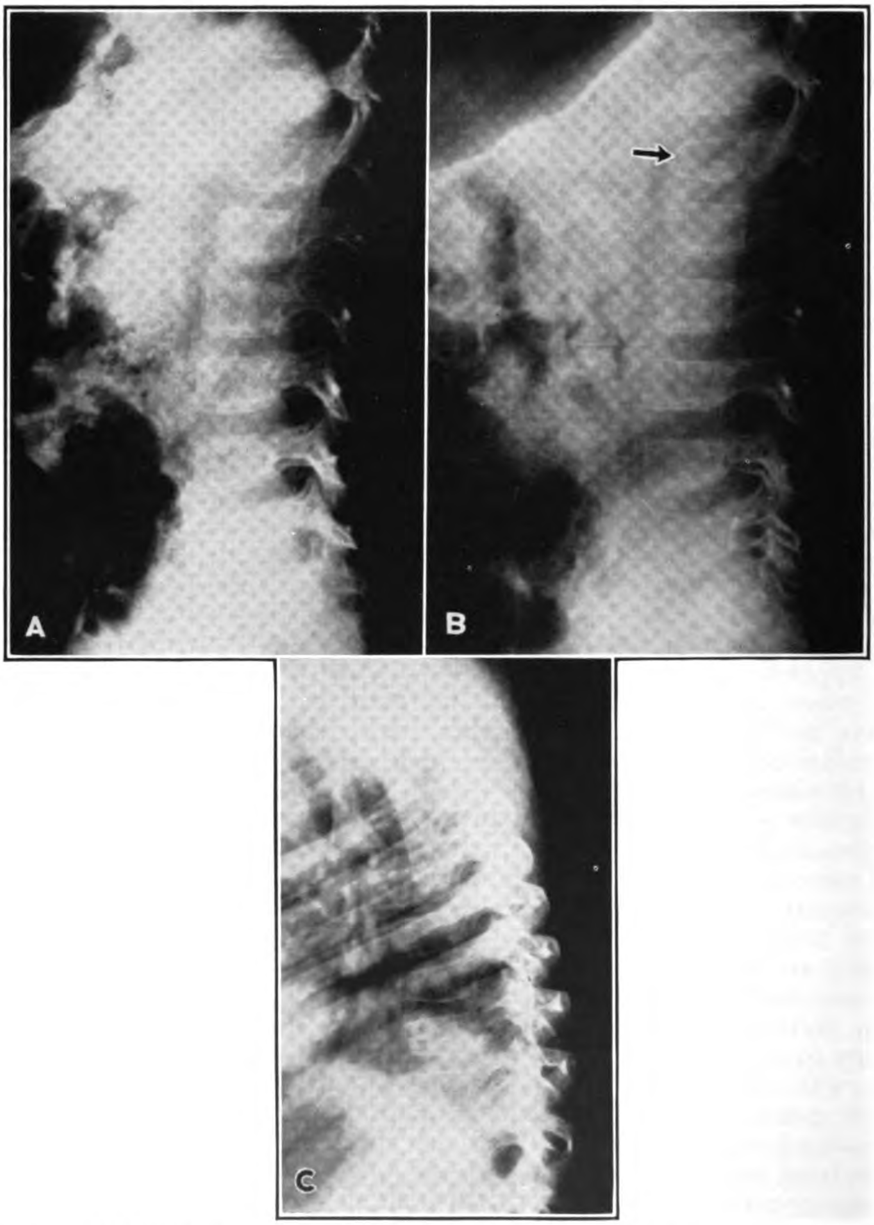

FIG. 370.—*A*, Acute leukemia in a four-year-old child. Marked demineralization of the entire spine is present. *B*, Same case, fifteen weeks later. Note the partial collapse of the anterior aspect of the body of the twelfth thoracic vertebra. The osteoporosis is more marked. *C*, Same case, thoracic vertebræ. Marked osteoporosis is present, with partial collapse of several of the midthoracic vertebral bodies.

order of frequency. Baty and Vogt (1935) mentioned the occurrence of leukemic destructive changes in the spine and pelvis in children with osteoporosis, softening and compression of the vertebral bodies. This was similar to that observed with lymphoblastoma and neuroblastoma. Hildebrand (1950) stated that in pronounced cases the vertebræ may assume a biconcave appearance. Otherwise, bone destruction is represented by osteoporosis or a wedge-shaped deformity of the involved vertebral bodies. These changes may precede clinical manifestations by several months. Uehlinger (1952) also observed osteoporosis and the biconvex appearance of the vertebræ, and regarded these changes as nonspecific. He further postulated that it not the tumor cells alone that caused the osseous alterations but rather their effects upon the osteoblasts and osteoclasts, with destruction of the former and sparing of the latter. He mentioned that infiltration of the bone marrow may be generalized, with the axial skeleton less often affected than the appendicular skeleton.

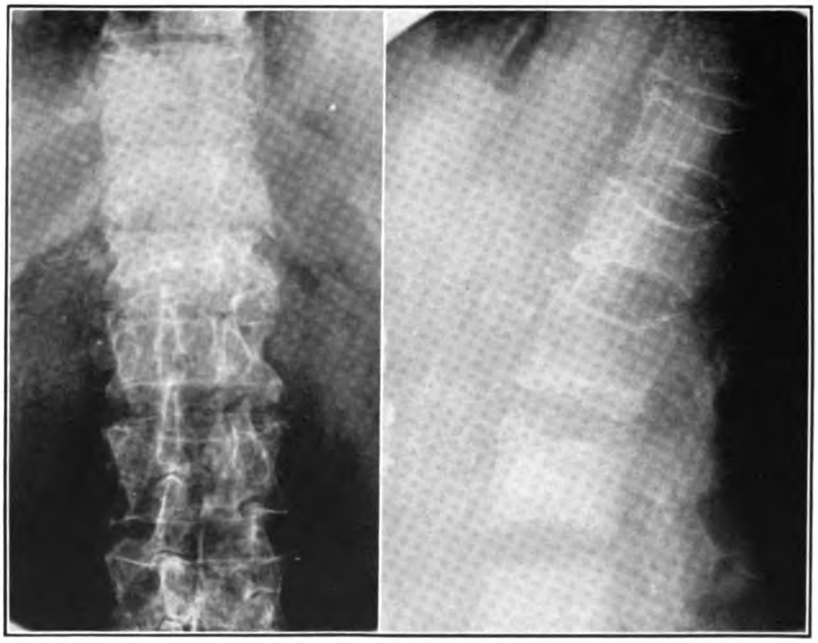

FIG. 371.—Myelogenous leukemia in a sixty-two-year-old woman. There is marked osteoporosis of the thoracolumbar spine, with compression of the first lumbar vertebra. The intervertebral discs are biconvex.

Jaffe (1952) mentioned that from 50 to 60 per cent of leukemic children revealed skeltal manifestations, and that the most frequent changes consisted of the narrowed transverse zone of radiolucency at the distal ends of the long bones. He, too, noted the occurrence of infractions and compression of the vertebræ. During remissions and while under treatment, the bony lesions may regress. The radiolucent zones are then replaced by dense bands such as may be seen with heavy metal poisoning. However, the original lesions reappear when the disease becomes reactivated with onset of the terminal phase. Skeletal changes were observed in adults in from 8 to 10 per cent of cases. These were principally osteoporosis, osteolytic foci, periosteal new bone formation and fractures.

Windholz and Foster (1949) observed roentgenologic evidence of monostotic or polyostotic sclerosis in leukemia. This had no bearing on the clinical course, and suggested that the bone marrow was becoming fibrous and aplastic. Osteosclerosis of this type was more common in monocytic leukemia and in aleukemia lymphadenosis. One case of lymphatic leukemia had panostotic sclerosis.

An unusual case of extradural leukemic implants was reported by Mosberg (1951) in a five-month-old child who became paraplegic within a period of four weeks. At laminectomy an extradural leukemic deposit was found in the thoracolumbar region. The plain films in this case showed dilatation of the spinal canal at the level of the twelfth thoracic and first lumbar vertebræ, and myelography revealed a complete block at the first lumbar interspace. This patient was reported as 1 of 23 cases of intraspinal tumors occurring during the first year of life, and Mosberg commented upon the high association of these growths with congenital anomalies.

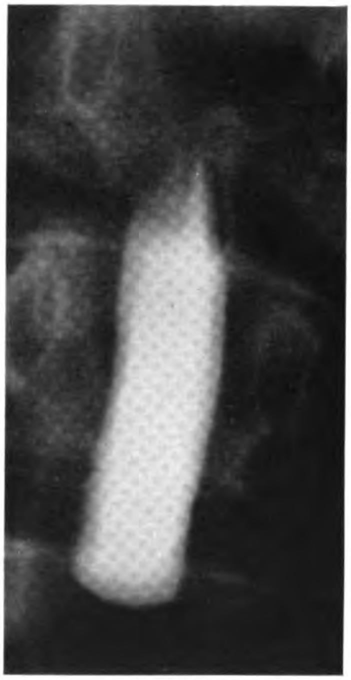

FIG. 372.—Epidural mass of leukemic tissue causing a complete block in the lower thoracic spinal canal in a 16-year-old male, verified at operation.

The radiologic changes produced by leukemia in the vertebral column are more frequent than generally recognized, especially in infants and children. The usual change observed in younger patients is a diffuse demineralization of bone, loss of trabecular markings and a biconvex appearance of the vertebræ. Anterior compression of the midthoracic vertebræ is relatively frequent. There also appears to be a tendency for the anterior and posterior venous grooves to be persistent and rather prominent. The use of steroid and chemotherapeutic measures influences the bony changes, and sometimes one cannot decide whether the bone loss is due to the leukemia or the effects of medication, especially steroids.

In older children of from about two to six years of age the vertebral bodies also become demineralized, but the loss of bone is more prominent in the centrum of the vertebral body and in the epiphyseal zones at the superior and inferior aspects of the involved vertebræ. Zones of increased radiolucency appear at the cartilaginous-osseous junctions, comparable to those seen so frequently at the diaphyseal ends of the growing long bones. Occasionally one can identify one or two parallel lines of increased density between the demineralized epiphyseal zones and the centrum. Frequently the vertebral bodies present rounded margins, or are diffusely and variably flattened.

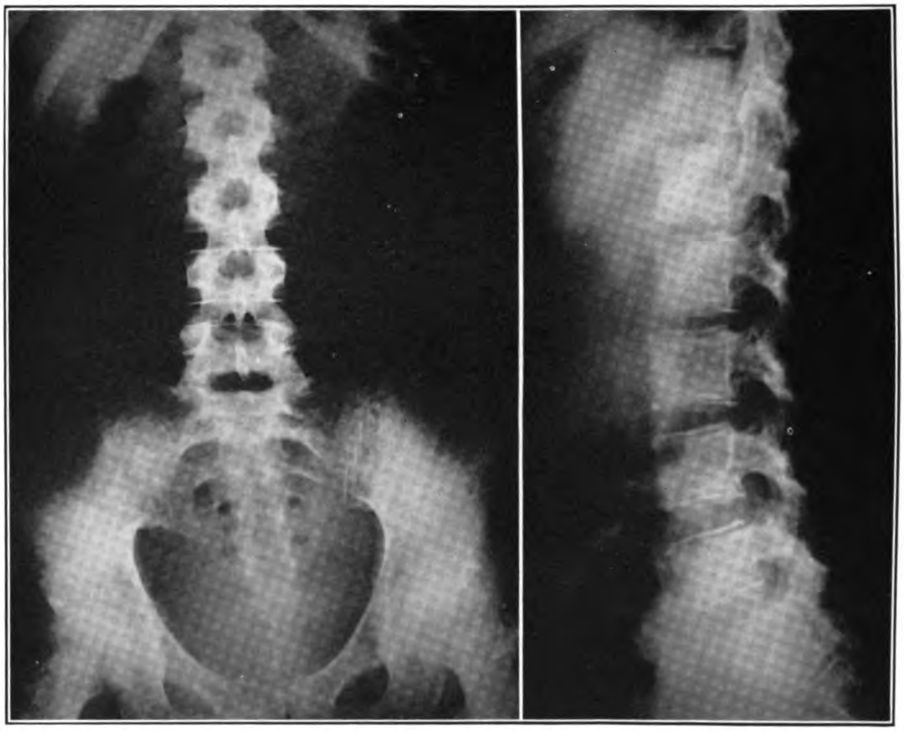

FIG. 373.—Leukomyelosarcoma in a thirteen-year-old girl. Marked osteoporosis of the vertebral bodies, with rarefied zones immediately above and beneath the articular plates. At necropsy there was a tremendous increase in the myelocytic elements in the marrow cavity infringing on the trabeculæ and causing a dissolution of their continuity, fragmentation, localized atrophy and osteoporosis. In some areas no bone was visible, the entire portion being taken up by the proliferating cells. The cortical bone was thinned out.

When sufficiently pronounced, hepatosplenomegaly causes a straightening of the usual lumbar lordotic curve. When these organs overlay the spine they tend to obscure osseous detail, contributing to the loss of trabecular markings. Occasionally retroperitoneal hemorrhage or ascites add to the difficulties in evaluating osseous patterns. The psoas margins may be obscured or obliterated, or the bladder deformed by retroperitoneal hemorrhage.

Changes in the growing ends of the vertebræ do not appear in patients past adolescence, but demineralization represents one of the more important changes incident to leukemia, especially in young adults. Occasionally leukemic infiltrations in adults behave as do other locally invasive tumors, causing bone destruction and consequent partial collapse. This is relatively infrequent, and cannot be distinguished from other metastatic lesions on the basis of radiologic changes alone. Osteoporosis in this group is likely to be exaggerated if they have been undergoing steroid treatment.

The clinical picture associated with vertebral involvement of the spine with leukemia usually is rather mild in contrast with the changes produced. Occasionally a child complains of back pain, but rarely is this intense. In adults with local bone destruction the symptoms are comparable with those due to similar lesions of non-leukemic origin.

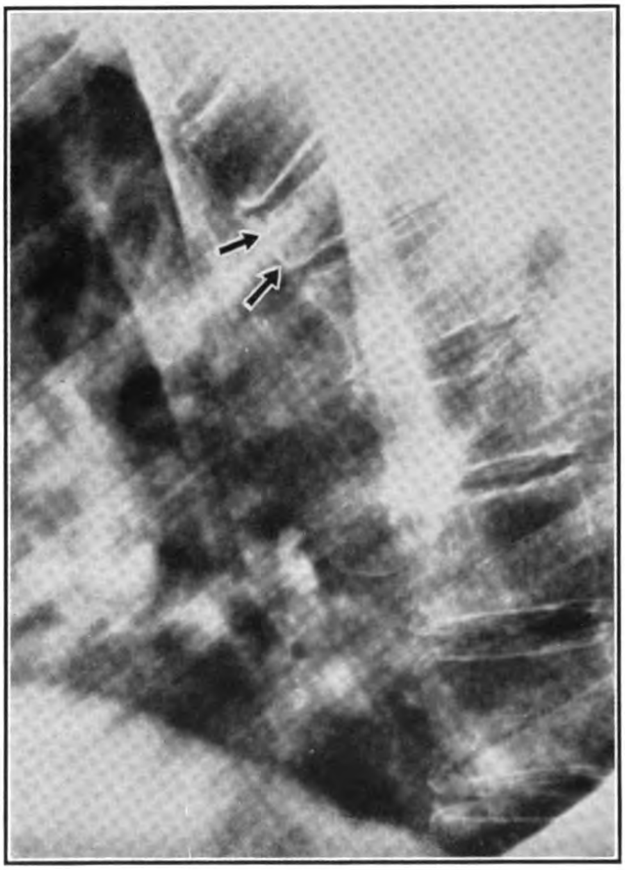

FIG. 374.—Acute myelogenous leukemia in a fifty-three-year-old man, with partial collapse of the fourth dorsal vertebra (arrows).

In our experience leukemic infiltration of the spinal meninges is not uncommon, but the production of clinical complaints because of such infiltrations is infrequent. Occasionally an epidural mass of leukemic tissue produces the usual picture of an intraspinal tumor, and this is best localized by means of myelography. The spinal nerve roots also may become infiltrated with leukemic deposits, with consequent neuritic pain. Where a tumorous deposit of leukemic tissue causes compression of the cord and cerebrospinal fluid block, prompt localization by myelography, followed by appropriate surgery and radiation therapy, may ameliorate the symptoms.

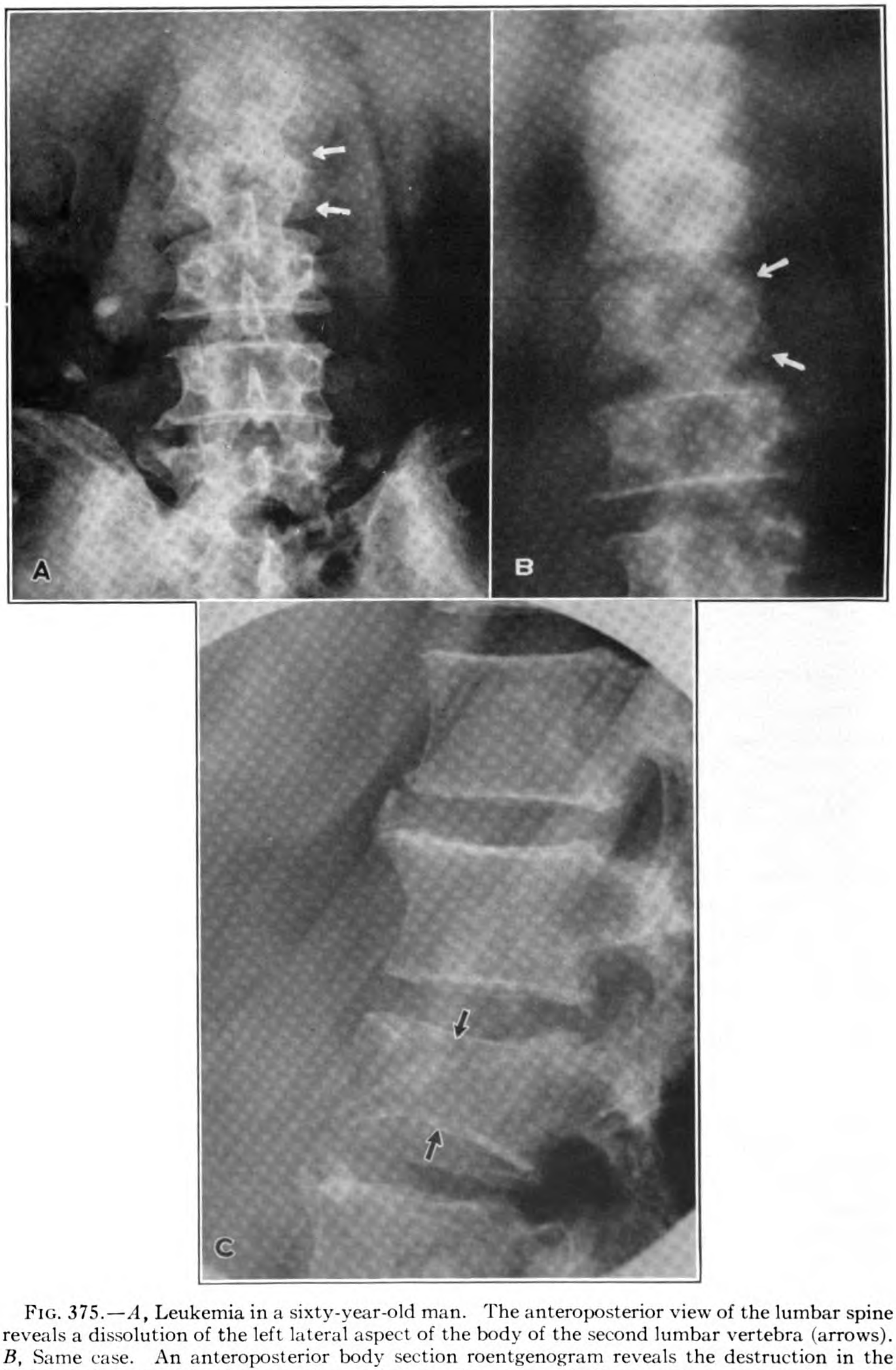

FIG. 375.—*A*, Leukemia in a sixty-year-old man. The anteroposterior view of the lumbar spine reveals a dissolution of the left lateral aspect of the body of the second lumbar vertebra (arrows). *B*, Same case. An anteroposterior body section roentgenogram reveals the destruction in the body of the second lumbar vertebra to better advantage. *C*, Same case, lateral lumbar spine roentgenogram showing partial collapse of the body of the second lumbar vertebra (arrows).

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MYELOSCLEROTIC ANEMIA

Various names have been applied to this disease, including myelosclerosis, myelofibrosis, leukoerythroblastic anemia, myelophthysic anemia, non-leukemic myelosis, agnogenic myeloid metaplasia, and agnogenic splenomegaly. The condition is characterized by generalized fibrosis of the bone marrow with excessive proliferation of endosteal bone. The fibrosis may involve the marrow cavity alone, or it may invade the bone and extend to and elevate the periosteum. The patches of fibrosis within the marrow cavity are scattered or confluent, and vary greatly. The entire skeleton may be involved, particularly the blood-forming marrow as represented by the spinal column, the pelvis, ribs and the proximal portions of the long bones. As a result of the replacement of the red marrow by fibrous tissue, compensatory hyperplasia appears in the remaining hematopoietic tissues, particularly the liver, the spleen, and occasionally in the ovaries and adrenals.

Examination of the circulating blood reveals immature red blood cells and leukocytes which may lead to confusion with chronic myeloid leukemia. Anemia is frequent, although in the early stages of the disease a moderate polycythemia may occur. Splenomegaly becomes pronounced later in the disease, and is one of the factors adding confusion to the differential diagnosis. According to Sussman (1947) the disease may be classified as an atypical form of myelogenous leukemia, or a megakaryocytic leukemia because of an increase in megakaryocytes in the bone marrow or other organs. While a moderate polycythemia is not uncommon in early stages, anemia in these patients finally becomes progressive. Leukoblastic activity is heightened and death may follow leukemic invasion of the spleen and other organs, or bone marrow exhaustion may occur with myelosclerosis. Windholz and Foster (1949) pointed out that either splenectomy or *x*-ray therapy might hasten death, commenting that many such patients may live comfortably for thirty years or more without treatment. Bastrup-Madsen (1952) emphasized the importance of differentiating myelosclerotic anemia from other diseases in which radiation therapy or splenectomy might be contemplated, inasmuch as these procedures might prove dangerous in myelosclerotic anemia. There is evidence, however, that *x*-ray therapy or splenectomy actually may be palliative when hypersplenism is pronounced (Linman and Bethell, 1957; Leigh, Corley, Huguley and Rogers, 1959).

The radiologic changes seen with myelosclerotic anemia, as indicated by the name itself, are predominantly of a sclerotic nature. In the vertebral column the splotchy confluent areas of increased density appear in the spongiosa as fairly uniformly distributed patches in the thoracic and lumbar vertebræ, and are less frequently seen in the cervical vertebræ. No alterations in the configurations of the vertebræ or the intervertebral discs have been reported. Less frequently the bony changes are diffuse, giving rise to a relatively increased density of the vertebræ with loss of definition of the trabecular markings and replacement of these by a diffuse, almost ground-glass increase in osseous density. The combination of hepatosplenomegaly with osteosclerotic changes in the vertebræ without alterations in their contours is regarded as a fairly reliable diagnostic point.

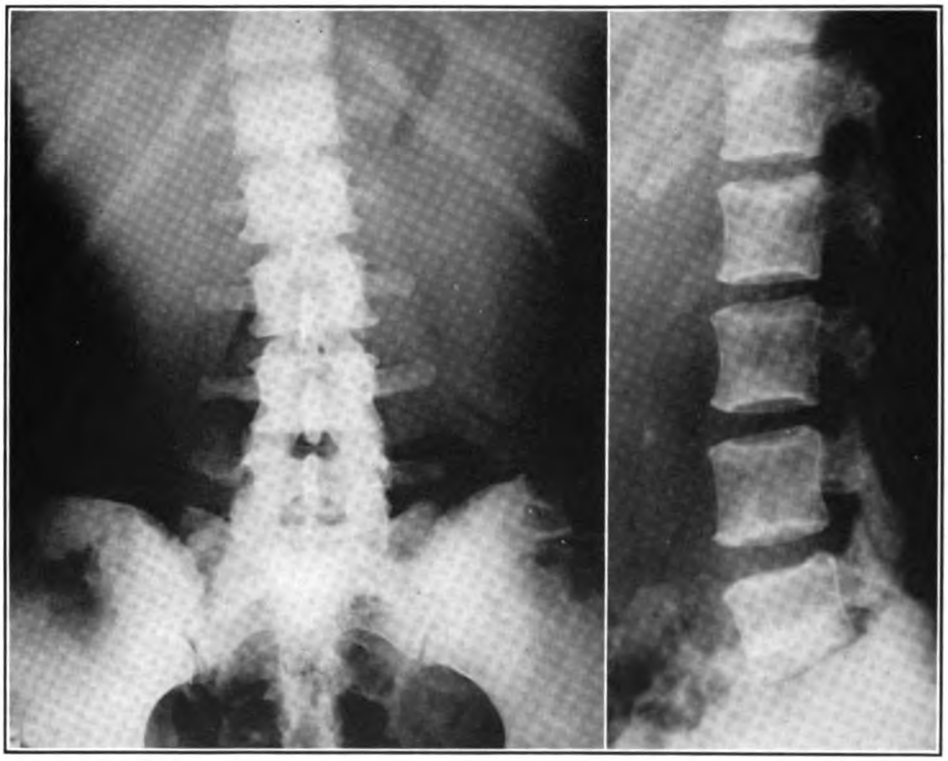

FIG. 376.—Osteosclerotic anemia in a forty-two-year-old woman who had the disease for thirteen years. There is marked hepatosplenomegaly. The lateral view shows straightening of the lumbar lordotic curve and diffuse mottling of the vertebral bodies.

Microscopic examination of the diseased tissue reveals a diffuse or focal proliferation of endosteal bone, with prominent, large active osteoblasts.

Osteoid tissue is minimal or absent, and spicules composed of osseous tissue may be found scattered through the medullary spaces. A distinct paucity of osteoclasts has been reported. This results in prominence and overgrowth of the trabeculæ which may become confluent in an irregular manner, producing scattered foci of denser bony spicules merging with the endosteum.

Secondary myelosclerosis also may occur with other diseases of the osseous system, of which metastatic prostatic carcinoma is the most common (Bersack and Feinstein, 1946). Diffuse endosteal sclerosis may also be seen with widespread metastases from carcinoma of the breast, stomach or colon. Reported elsewhere in the present monograph are 2 cases of carcinoma of the stomach with widespread myelofibrosis secondary to metastatic disease in which the primary lesion was disclosed only at necropsy.

Osteosclerosis also occurs, although infrequently, with longstanding chronic renal insufficiency, and affects the lumbar vertebræ, the pelvis, the sacrum and less often the chest, skull and femurs. The cause for this is unknown, and the change is in interesting contrast with the more frequently observed resorptive changes associated with the secondary hyperparathyroidism associated with uremia. The presence of osteosclerosis in patients with urticaria pigmentosa has been reported also. Collections of mast cells producing mixed osteoblastic and osteolytic reactions have been observed in the spine as well as in other bones. The absence of splenomegaly and the usual features of renal disease, and the skin changes of urticaria pigmentosa serve to distinguish these conditions clinically from agnogenic myeloid metaplasia.

Myelosclerosis may also occur in Paget's disease, myeloma, terminal polycythemia vera, tuberculosis and syphilis. Benzene, fluorine, lead, arsenic and phosphorus poisoning are also considered causative (Baastrup-Madsen, 1952). Similar changes may also be part of the picture of metabolic disturbances such as Albright's disease, osteomalacia, osteitis fibrosa cystica and the various lipid dyscrasias (Erf and Herbut, 1944). One of the conditions which is frequently included as a cause of osteosclerotic anemia is Albers-Schönberg's disease, but this represents a defect in development of mesenchymatous elements rather than a fibroblastic reaction to a stimulus which also produces extramedullary hematopoiesis.

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POLYCYTHEMIA VERA

This condition, characterized by a pronounced increase in the circulating red blood cells, is associated with cyanosis, particularly of the face and mucous membranes, enlargement of the liver and to a lesser degree of the spleen. Associated complaints are headaches, diffuse aches and pains over the extremities and trunk, easy fatiguability, dizziness, tinnitus, dyspnea on exertion and mental depression. The concomitance of polycythemia vera and peptic ulcer and various neurogenic diseases centering on the midbrain-pituitary area have been described. The association of polycythemia vera with leukemia is recognized, and it is today believed that the disease is one not limited to the erythropoietic system but rather involves the entire blood-forming apparatus. Polycythemia may be accompanied by a corresponding non-malignant proliferation in the leukoblastic portion of the bone marrow, as evidenced by the elevated white cell count not infrequently encountered. A high incidence of leukemia is noted in patients surviving more than ten years.

Histologic examination of sections of the bone marrow from the sternum show hyperplastic marrow with replacement of fatty tissue by hematopoietic tissue. Hyperplasia of cells of the megakaryocytic, erythroid and myeloid series and a tendency for the cells of the erythroblastic series to occur in small clusters was reported by Lawrence, Berline and Huff (1953). In 2 cases observed by them at necropsy hyperplastic bone marrow was found in the vertebræ. In 1935, Hirsch reported post-mortem observations in a male patient fifty-five years old who was known to have had polycythemia for thirty-one years. Examination of the bodies of the lumbar vertebræ showed the marrow to be tan-red in color and reduced to approximately one-fifth of its usual volume. The bone trabeculæ in the vertebræ were markedly thickened and the dimensions of the marrow spaces were correspondingly reduced. Along the edges of the trabeculæ were narrow layers of new bone. Hirsch concluded that the generalized osteosclerosis in this case was probably part of the picture of the advanced chronic stage of the disease. Stodtmeister and Sandkühler (1952) also mentioned that patients with polycythemia vera who survived long enough developed myelofibrosis which ultimately might progress to osteomyelosclerosis.

The radiologic changes in the spine associated with polycythemia vera are minimal. In a review of 88 proved cases Hodgson, Good and Hall (1946) were unable to find any bone lesions attributable to the disease. However, in cases of extremely long duration such as that described by Hirsch, radiographic examination of the spine might in occasional instances reveal sclerotic bone marrow changes.

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ACQUIRED HEMOLYTIC ANEMIA

This is a disease of unknown etiology in which the red blood cells are destroyed, presumably by an auto-agglutinin firmly attached to the red cells and capable of reacting with normal human erythrocytes. It may ap-

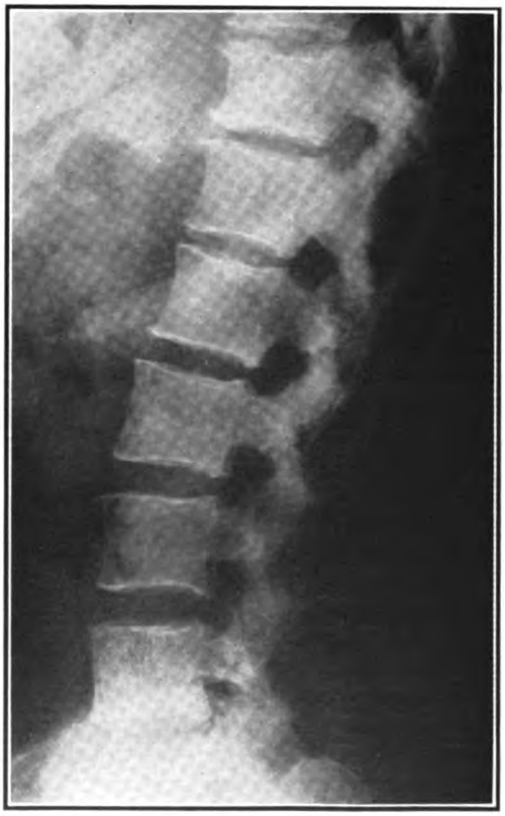

Fig. 377.—Lateral lumbar spine of a thirty-seven-year-old male who had had a splenectomy seven years before for acquired hemolytic anemia. There is a faint diffuse and spotty osteosclerosis in the vertebral bodies.

pear in previously healthy persons or may develop in patients with primary atypical pneumonia, with lymphomas or with collagen disease. It may occur at any age, but is more frequent in females. The condition was reviewed recently by Sacks, Workman and Jahn (1952) who reported on 19 patients studied by them and 47 collected from the literature. No mention was made of skeletal changes in this condition.

In our own material was 1 male patient thirty-seven years old who had had a splenectomy for acquired hemolytic anemia seven years before the present films were made. Spine roentgenograms revealed osteosclerotic changes, much the same in radiologic appearance as those seen with osteosclerotic anemia. Of interest in this particular patient is the fact that *x*-rays of the lower femurs showed a flask-like appearance similar to that described in Gaucher's disease. Mild osteosclerotic changes were also present in the distal ends of the femurs and the upper ends of the tibias.

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Sickle Cell Anemia

This condition is a hemolytic anemia occurring principally, but not exclusively in the Negro race. It is a chronic disease of a familial nature, and is characterized by the presence of sickle-shaped red blood cells. This defect has been attributed to a characteristic inherited type of hemoglobin which can be differentiated from normal by electrophoresis. The abnormal hemoglobin designated as Hemoglobin S is inherited as a dominant Mendelian characteristic. If this is received from both parents a homozygous state results in sickle cell anemia. When the S factor comes from but one parent a heterozygous state produces the sickle cell trait. Sickling then appears under stresses like exposure to cold, and symptoms appear only if thrombotic phenomena follow. Bony changes are rarely seen in patients with the sickle-cell trait. Another hemoglobin variant, hemoglobin C, also can interact with hemoglobin S to produce sickle cell hemoglobin C disease, which is milder than the true sickle cell homozygous S/S disease.

In acute stages of S/S sickle cell disease vascular occlusive phenomena affecting all the organs of the body produce variously abdominal, central nervous system, bone and joint, urogenital and skin lesions. Clinically there are periods of exacerbation and remission. Anemia, leg ulcers, abdominal and joint pains, splenomegaly, cardiac murmurs, fever and leukocytosis may occur in children. Bone and joint pains may predominate, and be sufficiently severe to simulate osteomyelitis. The joint pains may be migratory and associated with fever and leukocytosis, simulating rheumatic fever or rheumatoid arthritis. The widespread changes encountered in the various visceral organs are due to thromboses from the rigid sickle cells in the capillaries, which rapidly ascend to larger vessels, particularly on the arterial side. The crises of sickle cell anemia follow the widespread thromboses which appear as the capillaries become plugged. This may induce a widespread concomitant vasospasm. Hyperplasia of the bone marrow occurs in an effort to maintain the circulating blood. This takes place in the cancellous bone, and is most marked in the calvarium, the long bones and the axial skeleton.

Patients with sickle cell anemia of long duration present a fairly characteristic habitus in which the patient is underweight, thin, with narrow hips and

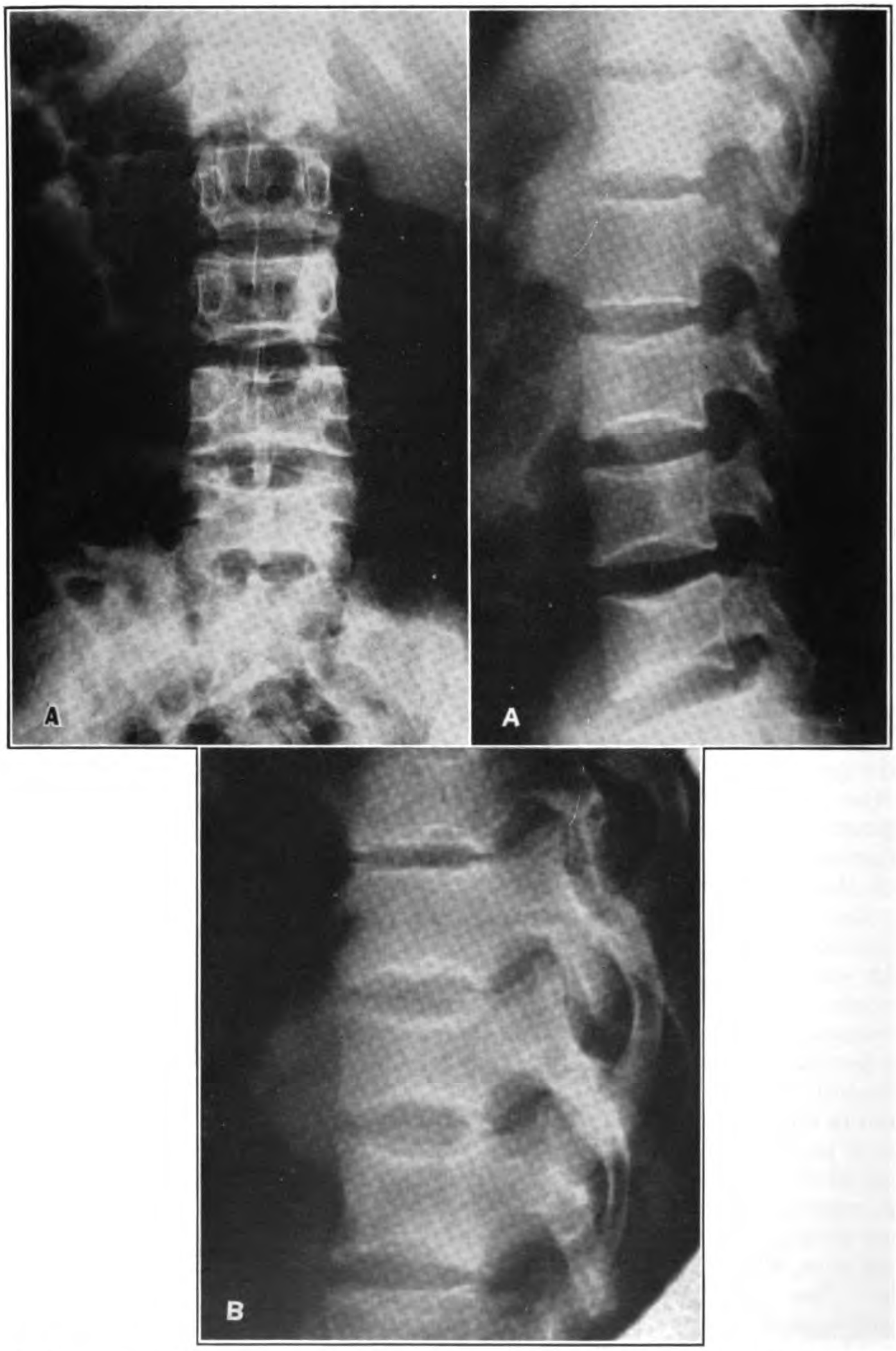

Fig. 378.—*A*, Sickle cell anemia in a nineteen-year-old male negro patient. The lumbar spine reveals straightening of the lordotic curve. The vertebral bodies are intact, but diffuse streaky increased density of the cancellous bone is present. *B*, Same case, lateral thoracic spine. The bony changes are more pronounced in this region.

shoulders and decreased stature. An increase in the upper dorsal spine curvature and lumbar lordosis is common. The neck and trunk are short, and an increase in the anteroposterior diameter of the chest is often present. The abdomen may protrude and the extremities remain quite thin. The hands are long, narrow and tapering. Margolies (1951) believed that the characteristic body habitus was probably due to anemia during the formative years, and to the frequent arteriolar thromboses. The decreased weight and stature was presumed to be due to changes in the anterior pituitary body, which in turn is accompanied by atrophy of the testes and other signs of hypogonadism in males. The effects of sickle cell anemia on the endocrine system, the cardiovascular system, the liver and spleen and the abdominal organs is beyond the scope of this monograph.

Changes in the skeletal system are observed fairly frequently, principally in the skull, the spinal column, the femurs and the tibias and fibulæ. Less commonly rarefied, trabeculated changes with patches of osteosclerosis appear in the small bones of the hands and feet and the long bones of the upper extremities. The bony changes are produced by hyperplasia of the erythroblastic tissues in the bone marrow. As the marrow is increased in bulk there is a concomitant attempt to replace the marrow with osteoid tissue and new bone. In childhood, the predominant change is usually widening of the medullary canals and thinning of the cortex of the tubular bones. In a case reported by Henkin (1949) with collapse of the vertebral bodies, examination of the bone marrow at necropsy in a fourteen-year-old Negro boy revealed great cellularity and congestion, with trabecular destruction and connective tissue replacement taking place almost simultaneously in the erythropoietic portions of the cancellous bone. He believed that the process was a form of osteoporosis in which the osteoblasts do not lay down sufficient osseous matrix because of a numerical decrease in osteoblasts rather than a decrease in their activity. This process is intermittent, and probably associated with diminished circulation due to the thrombotic tendencies affecting the intraosseous circulation. This combination of intraosseous destruction of trabeculæ, hyperplasia of the marrow and reparative processes is basically responsible for the presence of patchy areas of demineralization and increased bone activity.

The early radiologic changes in the spine associated with sickle cell anemia are a more or less diffuse osteoporosis with a biconvex appearance of the vertebral bodies and widening of the intervertebral discs. Later in the disease the vertebral bodies become flattened, and as reparative efforts occur, osteosclerotic foci become demonstrable as streaky interwoven filaments of increased bony density within the vertebral bodies. These changes occur either in the thoracic or in the lumbar vertebræ, the cervical spine usually remaining unchanged. In a report of 48 cases with examinations of the spine, Ehrenpreis and Schwinger (1952) reported that changes were found in 34. These were marked in 7 cases, moderate in 14 and minimal in the remaining 13.

The roentgenologic appearance of the vertebræ varies with the stage of the disease encountered at the time of examination. In young individuals in whom there has been relatively little in the way of occlusive phenomena and reparative effort, there may be no visible changes in the vertebral bodies at all. With marked hyperplasia, the only radiological indication may be some osteoporosis of the bodies, and widening of the intervertebral discs. When the occlusive phenomena are more advanced, and reparative efforts have taken place to an appreciable degree, the interlacing streaky appearance of the vertebral bodies are quite prominent. In the later stages, particularly in adults, the efforts at repair may be pronounced, and the vertebral bodies assume a biconvex appearance together with pronounced patchy sclerotic areas.

The occurrence of vertebral changes incident to what was presumed to be a massive infarction was reported by Legant and Ball (1948). In 1 of their patients, a thirty-nine-year-old male, the lowermost two lumbar vertebræ presented an unusual appearance. They noted that two weeks after an abdominal crisis roentgenograms of the spine were normal. Eight weeks later areas of rarefaction appeared, with compression of the adjacent margins of the fourth and fifth lumbar vertebræ. Ten weeks later the bodies were further compressed with areas of rarefaction in the anterosuperior aspect of the fifth lumbar vertebra. Eight, ten and twelve months thereafter there was restitution of the architecture of these vertebral bodies almost to normal, with a hemogeneous increase in the density of both vertebræ and narrowing of the third and fourth intervertebral discs. Actual collapse of the vertebral bodies with sickle cell anemia is rare. Carrol and Evans (1949) stated that this had not been reported in the literature up until the time of their report. They described an eleven-year-old patient in whom there was destruction and collapse of the bodies of the ninth and tenth thoracic vertebræ.

Our experience with sickle cell anemia corresponds to that described above. The youngest case observed by us with spine changes occurred in a three and one-half-year-old male patient in whom the vertebral bodies revealed a mild osteoporosis associated with thin fine lines of increased bone density, particularly in the bodies of the third, fourth and fifth lumbar vertebræ. The intervertebral discs in this patient were not disturbed, nor was there vertebral collapse. Along the anterior aspects of the bodies of the lowermost four dorsal vertebræ small, almost rectangular tongue-like protrusions were observed.

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THALASSAEMIA

In 1927 Cooley, Witwer and Lee reported an unusual type of progressive anemia in childhood with erythroblastosis, splenomegaly, hepatomegaly and peculiar osseous changes. This is also called Mediterranean anemia, erythroblastic anemia, thallassemia, or target cell anemia. The patient customarily is underdeveloped, pale and listless and the condition also is associated with cardiac dilatation, edema and effusion into the serous cavities. The disease is familial, and while generally observed in people of Mediterranean origin, has been reported in others such as Chinese, Mexican, American Indian, Filipino and other non-Mediterranean people (Silver, 1950; Minnich, Na-Nakorn, Chong-Chaveonsuk and Kochasini, 1954). Cooley's anemia may affect adolescents and adults in a less severe form, with the patient completely unaware of his illness. The clinical differentiation between Cooley's anemia and Cooley's trait and their genetic relationships were discussed by Daland and Strauss (1948). The thalassaemia gene also can combine with that of the sickle cell trait, resulting in sickle cell thalassaemia disease (MacIver, Went and Cruickshank, 1958).

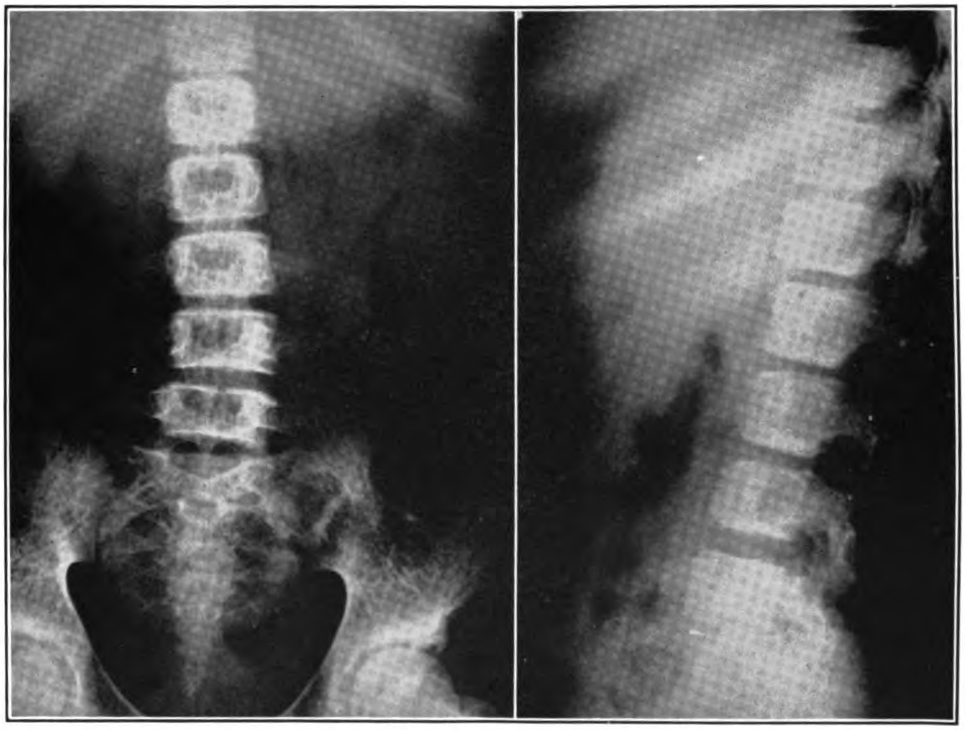

FIG. 379.—Cooley's anemia in an eleven-year-old girl. There is marked reticulation of the vertebral bodies, the pedicles and laminæ and of the sacrum and iliac bones.

The milder forms of the disease hematologically may be associated with a mild or even absent anemia, increased reticulocytosis, absence of erythroblastosis and the presence of anisocytosis, poikilocytosis, target cells and basophilic stippling of the red cells in the peripheral blood. In the severe form the anemia may be pronounced, with hypochromic microcytic and nucleated red cells prominent in the peripheral smear. The blood shows signs of increased red blood cell formation with anisocytosis and poikilocytosis. In young children the leukocytic count may be increased, with an increased lymphocyte content. Myeloblasts and myelocytes may also be found. Aspiration of the bone marrow reveals marked hyperplasia of the red cells with a pronounced increase in normoblasts. The red cells are remarkably resistant to rupture in hypotonic saline solutions.

The osseous changes with thalassemia are due primarily to over-activity and overgrowth of the bone marrow. Consequently the changes in infants and in children occur practically in the entire skeleton, reflecting the disposition of red marrow. The characteristic osteoporosis, thinning of the cortex of the long bones, widening of the medullary canals and the traversing cancellous interlacing web-like strands of increased density may be observed in all the bones except the skull, which has its own peculiar hair-on-end picture. In patients surviving adolescence, regression of the bony changes may take place, particularly in the shafts of the long bones. Caffey (1951) followed 4 patients to their twenty-eighth, twenty-sixth, eighteenth and fourteenth birthdays, respectively, and found that after puberty the optimal sites for demonstrating osseous x-ray changes were the skull, the spine and the pelvis. This reflected the sites of active bone marrow as the individuals grew into adult life. Caminopetros (1938) reported bone changes of erythroblastic anemia in apparently healthy parents of

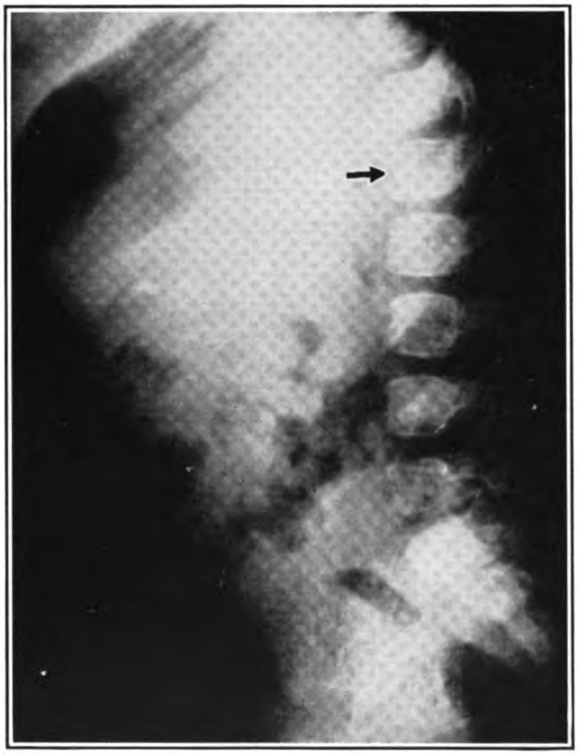

FIG. 380.—Cooley's anemia in a two-year-old girl. Note the osteoporotic and reticulated appearance of the vertebral column. There is a small, semicircular notch in the anterior aspect of the body of the first lumbar vertebra (arrow) at the entrance of the anterior external vertebræ vein

patients with this condition. March, Schlyen and Schwartz (1952) followed several patients in the fourth and fifth decades in the same family with characteristic changes in the bones including the vertebræ.

In a patient observed by us from her eleventh to sixteenth birthday, the skeletal changes persisted during the entire period. At necropsy she had hyperplasia of the entire bone marrow and lymphadenoid system, advanced splenomegaly and hepatomegaly, dilatation of the heart, pulmonary edema and atelectasis. The onset of the disease in this patient had been noted when she was four years old, and its gradual progression was associated with lassitude, weakness, a mongoloid facial appearance, coarse hair, a fullness of the head anteriorly, and a tremendous splenomegaly.

Roentgenograms of the spine in this patient were made over a five-year period, and showed the vertebral bodies to be of average size, with a slight bulging of their contours. No evidence of collapse or other deformity was seen. Diffuse coarse reticulation in the vertebræ involved the pedicles, the laminæ and the transverse and spinous processes. No deformity of the neural arches or intervertebral foramina were noted. Over the years the vertebral bodies grew normally in size and appearance, but no gross change occurred in the reticulations described above. Similar changes were present throughout the skeleton. In another patient sixteen months of age who had a complete skeletal survey, characteristic changes were present in the entire skeleton. It was of interest that the anterior aspects of the bodies of the lower three dorsal vertebræ and the upper two lumbar vertebræ showed an indentation which was roughly semicircular in configuration and corresponded in position to the entrance of the anterior spinal vein. A similar change of lesser degree was also present in 2 cases of sickle cell anemia. A third patient eight years of age, who had been under observation for seven years, showed similar changes in the spine.

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HEMOPHILIA

Hemophilia is a hereditary disease which is predominant in the male and recessive in the female. It does not occur in women, but is transmitted through them to their male offspring. The hereditary defects apparently involve the thromboplastic activity of the plasma, thereby preventing normal clotting. The disease is characterized by hemorrhage which may either follow trauma or be spontaneous. The joints most frequently involved are those of the extremities, particularly the hips, knees and elbows. Characteristic defects with areas of destruction of bone due to necrosis following hemorrhage are quite frequent. Subperiosteal hemorrhages and resultant proliferative changes together with erosion of the underlying cortex have been reported by Echternacht (1943), who remarked that a hematoma, when organized and calcified, might produce an *x*-ray appearance resembling bone sarcoma. An excellent clinical review was published by Brinkhous (1954).

Although considerable has been written concerning the bony changes in the appendicular skeleton, there is very little concerning changes which may appear in the spine with hemophilia. Priest (1935) reported a case of spinal cord compression caused by epidural hemorrhage due to hemophilia. Changes in the sacrum in 1 case out of 44 were mentioned by Ghormley and Clegg (1948). In 7 cases seen by us the spinal roentgenograms were all normal.

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LUPUS ERYTHEMATOSUS

This is one of the collagen diseases which as a group involve essentially the various connective tissues of the body. It occurs most often in young women, and is usually associated with a butterfly erythematous rash over the bridge of the nose and malar eminences, fever of an intermittent nature and involvement of the synovial and serous membranes producing pericarditis, pleuritis and polyarthritis, and altered bone marrow activity. In later stages evidence of the effects of vascular alterations in the skin, kidneys and other parenchymatous organs are manifested. The disease is protean in its manifestations, and may occur in the absence of any of the symptoms and signs listed above.

The neurological aspects of lupus erythematosus were reported by Sedgwick and von Hagen (1948). They believed that all the symptoms referable to the central nervous system might be explained directly by focal vascular disease resulting in hemorrhage or occlusion. Indirect central nervous system effects occur by reason of involvement of the kidneys or by hemorrhage due to thrombopenia. Piper (1953) reviewed the literature on central nervous system involvement in disseminated lupus, and found no cases of focal involvement of the spinal cord. He reported a case in a nineteen-year-old girl with vascular changes including thrombosis of the meningeal vessels resulting in ischemic necrosis of the lumbar cord. The patient had complained of numbness and tingling of the lower extremities which gradually extended upwards, with progressive weakness, marked diminution in motility and difficulty in urination. At necropsy focal swelling with infarction of the lumbar spinal cord was evident on reflection of the dura. This was due to vasculitis, with thrombosis of the vessels supplying the meningeal vessels to the lumbar spinal cord. Other cases have been reported in which the essential change was vasculitis with fibrinoid change in the involved vessels and an associated myelomalacia which might have given rise to an adhesive meningitis or chronic meningo-encephalitis.

We had the opportunity of observing a nineteen-year-old girl with all the clinical manifestations of lupus erythematosus who complained of pains in the mid-dorsal region associated with sensory disturbances referable to the same area. On myelographic investigation a hesitation in the passage of pantopaque was noted at the level of the seventh dorsal vertebra. This was rapidly followed by paraplegia and she presented a picture considered indicative of a transection of the spinal cord due to a vascular accident. Laminectomy was performed (Dr. A. J. Berman), and no mass lesion was encountered. Even though the coverings appeared to be normal, definite epidural venous dilatation was encountered. When the dura was opened the cord appeared normal. Although this cannot be considered a confirmed case the clinical history, and neurological changes suggest that it might be included under the present category.

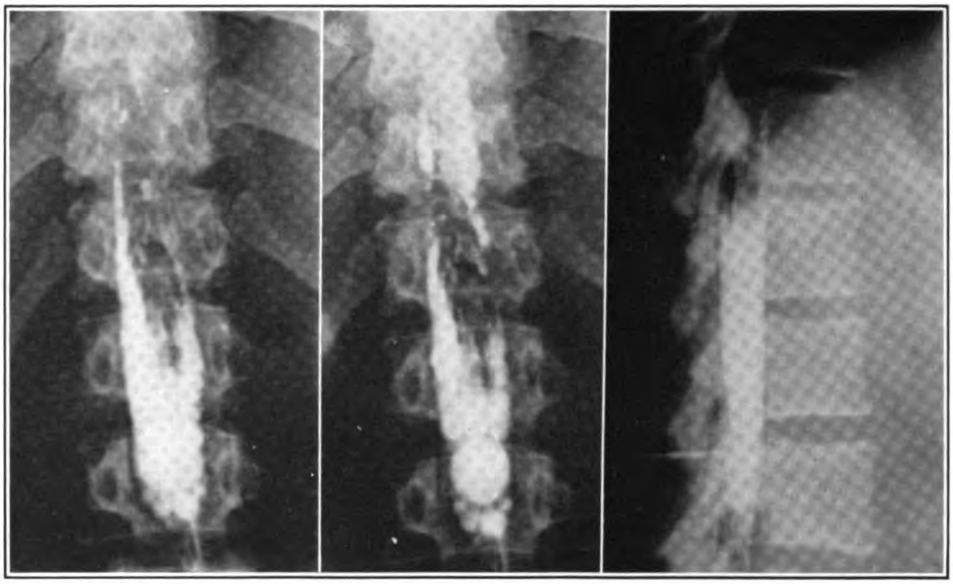

FIG. 381.—Myelograms of a nineteen-year-old girl with lupus erythematosus and paraplegia. A constant defect was present at the twelfth thoracic interspace. On the lateral view there was linear streaking in the pantopaque column below the defect believed due to edema of the nerve roots of the cauda equina.

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THE RETICULOENDOTHELIOSES

Gaucher's disease is one of the storage diseases of lipoid metabolism which involve particularly the reticulo-endothelial system. This condition, which is preponderant in females, is primarily a disturbance in kerasin metabolism in which deposition of kerasin occurs in the reticulum cells of the reticulo-endothelial system, producing a typical mottled infiltration with the foamy, so-called Gaucher's cells. The disease is a simple dominant hereditary one which is especially frequent in persons of Jewish origin, but may also occur in others. The clinical picture is varied, and may extend from childhood to old age. Symptoms appear as a rule during childhood and the progression of the disease may be slow, extending over many years.

Among the outstanding characteristics associated with Gaucher's disease are marked enlargement of the spleen and liver. The pronounced splenomegaly is responsible for the designation of the disease sometimes as splenic anemia. Involvement of the abdominal and intrathoracic lymph nodes and of the external lymph nodes is moderate or minimal. Brownish pigmentation of the exposed parts of the skin, wedge-shaped pingueculæ, a hemorrhagic tendency with leukopenia and thrombocytopenia may appear. It is customary to divide the condition into two forms designated as visceral and osseous types. In the first, enlargement of the spleen and liver and sometimes of the lymph nodes predominate, with relatively mild osseous involvement. In the osseous form the involvement of the skeleton is more marked, and the visceral changes are less pronounced. Anemia is usually more profound with the osseous form (Tennent, 1945).

The roentgenological changes in the skeleton associated with Gaucher's disease has been attributed to infiltration and replacement of the bone trabeculæ by the kerasin-containing reticulum cells. Infiltration of these cells around the small arteries and capillaries may be sufficient to produce aseptic necrosis (Schein and Arkin, 1942; Arkin and Schein, 1948). As a result of the infiltration of the marrow with Gaucher's cells minimal, moderate or extensive osteolysis may occur. Secondary changes such as bowing osseous collapse and pathologic fracture are not uncommon. In cases with bone involvement pain, stiffness and limping were the outstanding complaints in 11 cases reported by Welt, Rosenthal and Oppenheimer (1929). In the acute stages swelling, bone tenderness, and occasionally redness and limitation of motion suggested the diagnosis of osteomyelitis.

The bones most frequently affected by Gaucher's disease include the long bones, particularly the humeri and the distal ends of the femurs, the spine, pelvis and hips and those of the hands. Sections through these bones reveal the spongiosa infiltrated with yellow or grey deposits of Gaucher's tissue infiltrating between and replacing the trabeculæ. This is followed by thinning of the cortex from within, resulting in a scalloped, irregularly widened medullary canal.

Involvement of the spine with Gaucher's disease is by infiltration of the spongiosa with Gaucher's cells. As a result the vertebral body is weakened and wedge-shaped deformities appear. If the disease is extensive diffuse collapse of the vertebral bodies eventually results in flattening of the involved segments. Reed and Sosman (1942) described areas of rarefaction intermingled with areas of sclerosis in the involved vertebral bodies, followed by compression and relatively intact intervertebral discs. Kyphotic deformities with Gaucher's disease were mentioned by Tennent (1945). Welt, Rosenthal and Oppenheimer (1929) mentioned that 1 of their cases presented a gibbus without deformity of the intervertebral discs. Schein and Arkin (1942) in their eighth case reported numerous collapsed vertebræ which at necropsy were infiltrated with Gaucher's cells.

Windholz and Foster (1948) confirmed Pick's (1927) observation of sclerotic changes in the medullary canals of bones with Gaucher's disease, similar in appearance to regenerative ossification seen in inflammatory lesions. While the formation of osseous tissue as a rule was only slight, it pointed nevertheless to an effort at repair, and occurred in parts of the medullary canal without bone destruction as well as in structures adjoining typical absorptive changes. They believed that roentgen signs of pathological ossification in Gaucher's disease may be of diagnostic significance, and pointed out that such ossification appeared after splenectomy.

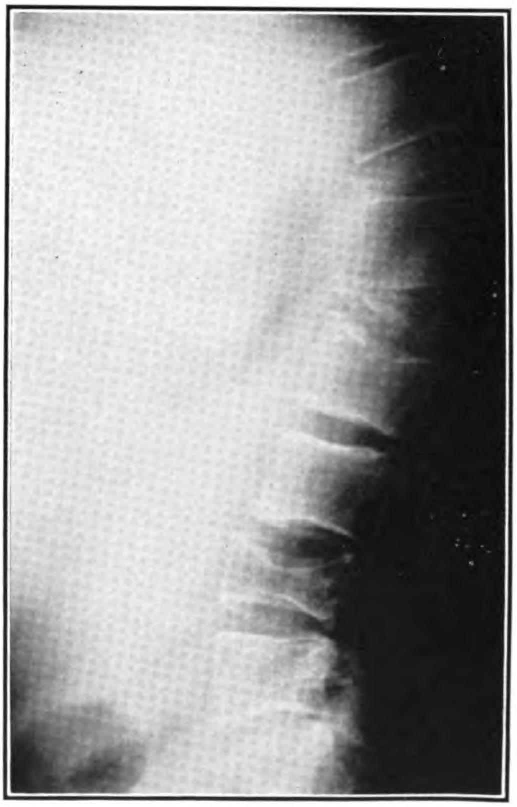

FIG. 382.—Gaucher's disease in a twenty-six-year-old male with multiple collapsed vertebræ.

Included in our own material are 7 cases of Gaucher's disease with roentgenograms of the spine. The youngest patient was four years old, with an advanced hepatosplenomegaly and early changes in the long bones of the extremities. In this patient only a mild degree of osteoporosis of the lumbar vertebræ could be discerned. The intervertebral spaces were normal, and the usual lumbar lordotic curve was straightened, probably as a result of the hepatosplenomegaly. It should be mentioned that the marked increase in the soft tissue densities overlying the abdominal area resulted in a peculiar haziness of the lumbar vertebræ as seen on the anteroposterior films.

Another youngster, twelve years old, showed no definite deformities of the vertebral bodies. One twenty-six-year-old patient was later reported in detail by Melamed and Chester (1938). The involvement of his spine was most extensive, and it was during our period of observation that his height diminished perceptibly. The appearance of the spine was such that we described it as an "accordion spine." Wedge-like deformities of the vertebral bodies of both the thoracic and lumbar spine first developed and later the vertebral bodies were reduced to thin discs no more than half a centimeter in height. The earliest change noted was a collapse of the superior aspect of the body of the first lumbar vertebra, with a tendency towards anterior angulation of the twelfth dorsal vertebra. The inferior aspect of the body of the first lumbar vertebra remained normal, and the intervertebral spaces above and beneath were not disturbed. Some increased density in the superior half of the compressed body of the first lumbar vertebra was observed initially, in all probablity due to compression of bony structure weakened by infiltration with Gaucher's cells. This patient, who was first observed by us in 1928, when the film mentioned above was

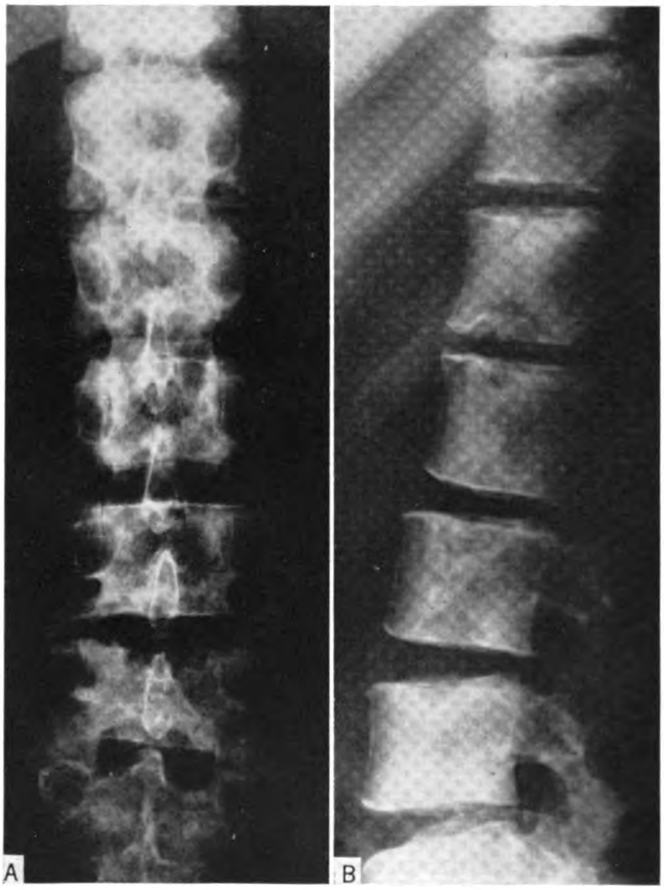

FIG. 383.—Gaucher's disease in a 42-year-old woman with diffuse vertebral sclerosis involving mainly the cancellous bone. The cortical bone is thin.

obtained, required splenectomy. He was again seen in 1932 with symptoms referable to multiple collapsed vertebræ and fracture of the sternum. At this time an enormous enlargement of the liver had appeared. The patient became mentally retarded and a generalized lymphadenopathy was found. The necropsy observations were reported in detail by Melamed and Chester.

Another woman, forty-two years old, had a splenomegaly which was first noted four years before she was seen by us. The diagnosis of Gaucher's disease was confirmed by positive splenic and bone marrow punctures. Her spine presented a most interesting appearance. The vertebral bodies were intact, but indentations of nuclear material into the bodies of the first and second lumbar vertebræ were seen on the lateral views. Slight narrowing of the intervertebral disc between these vertebræ was present. Particularly interesting was the presence of intermingled linear and slightly broader areas of bony increased density, indicating sclerosis in the spongiosa of the vertebral bodies. Sclerotic changes could also be demonstrated in the lateral aspects of the sacroiliac joints. This appearance was much the same as mentioned by Kulowski (1950), who described juxta-articular mottling in the sacroiliac joints with a tendency towards partial or complete obliteration of the joint spaces. A skeletal survey in this patient revealed characteristic changes of the Erlenmeyer flask deformity in the lower femurs. Moreover, extensive sclerotic changes were present in both lower femurs and in the humeri. Similar changes of lesser degree were seen in the tibias and in the dorsal spine.

Included among the lipoid granulomas are Niemann-Pick's disease and xanthomatosis. Niemann-Pick's disease is a storage disturbance of phospholipids, and bony changes are rare, although patchy osteoporotic areas may appear. Xanthomatosis occurs sometimes in a symptomatic form with cholesterolemia, in which lipoid granulomas of foam cells filled with cholesterol are deposited in tendon sheaths, joints or viscera. Bony involvement is rare.

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THE NON-LIPID RETICULOENDOTHELIOSES

This group includes Schüller-Christian's disease, eosinophilic granuloma of bone and acute disseminated reticuloendotheliosis (Letterer-Siwe's disease). Even though the clinical manifestations of these conditions are quite different, histologic evidence suggests a common origin for all three.

The progress of one of the three into one or both of the remaining two has been cited as evidence for the underlying similarity. However, Otani (1957) and Siwe (1949) disagree with this concept, and consider them separate entities because of dissimilarities in their clinical and pathologic aspects.

HAND-SCHÜLLER-CHRISTIAN DISEASE

This disease is characterized histologically by the presence of granulomatous tissue made up of large numbers of mononuclear cells, probably histiocytes, and a smaller number of leukocytes. Eosinophiles are present to a lesser degree. The cytoplasm of the histiocytes is finely or coarsely vacuolated. The granulomatous tissue contains varying amounts of foam cells and foreign body giant cells. These are more commonly observed in the spleen and lymph nodes than in the skeleton. The presence of foam cells may be minimal or absent in some cases, but in others they may be widely and extensively present (Ponseti, 1948).

With Hand-Schüller-Christian disease there may be storage of cholesterol in the foam cells which later is replaced by spindle cells and connective tissue. In older lesions, therefore, foam cells may be scant. The degree of eosinophilic infiltration may also vary considerably. The opinion has been expressed that foam cells are more commonly observed in the early stages of the disease, and disappear during the healing stage.

The triad usually associated with Hand-Schüller-Christian disease includes exophthalmos, diabetes insipidus and defects in the membranous bones. Concomitant with these are varying degrees of splenomegaly, pigmentation of the skin and evidences of endocrine dysfunction such as dystrophia adiposa genitalis and dwarfism. Polyuria, skin lesions and pulmonary manifestations may sometimes be present (Currens and Popp, 1943; Hodgson, Kennedy and Camp, 1951).

The bony lesions are most extensive in the skull, the pelvis and the extremities. The lesions usually are circumscribed, rarefied and may present scalloped borders, sometimes with increased density of the margins of the scallops. Involvement of the vertebral column is infrequent. Troxler and Niemetz (1946) reported a case of a thirty-five-year-old man in whom an area of destruction involving the anterior aspect of the body of the first lumbar vertebra was present. Davies (1949) mentioned a female child, two-and-a-half years of age, in whom there was a flattening of the body of the ninth dorsal vertebra. Lateral body section roentgenograms of the cervical spine in this patient showed considerable destruction of the body of the fourth cervical vertebra and loss of the joint space between the fourth and fifth cervical segments. Further survey of the skeleton showed lesions in the pelvis and femurs, and a biopsy from the left iliac crest was reported as Hand-Schüller-Christian type of reticulosis. Hodgson, Kennedy and Camp (1951) mentioned that the vertebræ were involved in 3 out of 28 cases of reticulo-endotheliosis, including in their series patients with Hand-Schüller-Christian disease, eosinophilic granuloma and Letterer-Siwe's disease.

EOSINOPHILIC GRANULOMA

This condition, together with Hand-Schüller-Christian disease and Letterer-Siwe's disease, is considered a nonlipoid reticulo-endotheliosis. The condition was first reported by Otani and Ehrlich (1940), who emphasized its granulomatous and benign nature, and the prominent eosinophilic

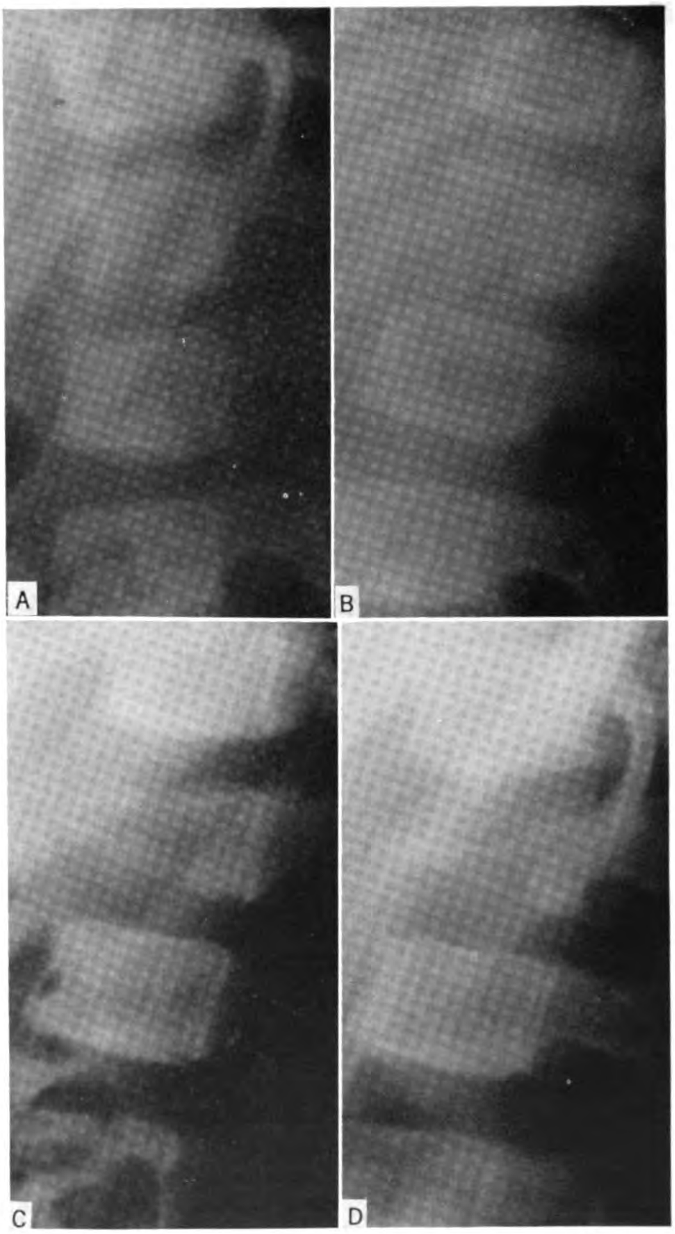

FIG. 384.—Eosinophilic granuloma of the first lumbar vertebra in a five-year-old boy. The initial complaint was moderate back pain which increased in severity. The films were made at intervals of four days. The rapid collapse of the vertebral body following diffuse loss of bone is well illustrated. In *A* there is demineralization of the anterior aspect of the vertebral body, which is more pronounced four days later (*B*). The intervertebral spaces above and beneath the involved vertebræ are widened as the vertebral body becomes flattened (*C* and *D*). Note how the posterior aspect of the vertebral body retains its height.

ing [illegible] bone. In their 7 cases, Otani and Ehrlich proposed ary anuloma of bone. Jaffe and Lichtenstein (1944) esion as being radiolucent and sharply delimited. On pathogic study in an early phase, the lesion appeared grossly as a more or less hemorrhagic and cystic area from which only a relatively small smount of granulation tissue could be curetted. The tissue was not necrotic, and contained conspicuous numbers of histiocytes. Some showed phagocytic activity, and interspersed among these were large accumulations of eosinophiles.

The disease usually occurs in young people. Green and Farber (1942) observed that their 10 patients varied in age from eleven months to ten years, eleven months. Gross and Jacox (1942), in a review of 60 cases, noted that all except three were younger than twenty-one years of age. One patient seen by us was a male fifty-eight years of age. Pain over the involved areas may be a prominent symptom. The regions involved most often include the skull, ribs and the long bones. Involvement of the spinal column is not uncommon. Green and Farber described a case in their series in which the second and third lumbar vertebræ were involved, associated with a paravertebral soft tissue swelling suggesting a tuberculous lesion. Another case showed a flattening of the ninth thoracic vertebra which remained unchanged over a four-year period. Dundon, Williams and Laipply (1946) mentioned that out of 46 cases collected by them from the literature, including 5 of their own, there were 10 cases with multiple lesions, of which 9 had vertebral involvement. In no instance of eosinophilic granuloma did vertebral involvement occur as an isolated lesion. Ackerman (1947) reported a nineteen-year-old male in whom there was a large area of destruction in the second cervical vertebra. The eleventh and twelfth dorsal vertebræ and all the lumbar vertebræ were markedly altered. The normal trabecular bony architecture was replaced by cystic areas of rarefaction involving the vertebral bodies and their processes. In addition there was some lateral wedging of the vertebræ. This patient also had involvement of the lungs and diaphragm with the eosinophilic granuloma. Walker (1952) mentioned a case involving the third lumbar vertebra with a destructive process, with recalcification of the vertebræ after *x*-ray therapy. Osborne, Freis and Levin (1944) reported a case in a twenty-one-year-old patient with evidence of neurologic impairment. This patient had headache, giddiness, vomiting, deafness, tinnitus, vertigo and ear pain as his chief complaints. There was involvement of the skull by a solitary confluent polycystic lesion in the temporal bone and mandible. Of interest to us was the fact that the seventh cervical, the first and second thoracic and the fifth lumbar vertebræ showed evidence of destruction. There was a favorable response to *x*-ray therapy in this patient. A case described by Weinstein, Francis and Sprofkin (1947) with headache associated with defects in the skull likewise complained of backache. Radiographic examination of the spine disclosed areas of destruction in the left iliac and pubic bones and the body of the third lumbar vertebra. This patient also presented radiological evidence of pulmonary fine nodular infiltrations, and proof of the eosinophilic granulomatous nature of the lesion was obtained by biopsy from the involved left parietal bone. This patient also did quite well with *x*-ray therapy.

Involvement of the spine is considerably more common with eosinophilic granuloma than with Hand-Schüller-Christian disease or Letterer-Siwe's disease. Of the 8 cases of Hand-Schüller-Christian disease and 5 cases of eosinophilic granuloma seen by us, two of the latter had vertebral involve-

ment. Both were identified because of flattened r [illegible] le
One was verified by vertebral biopsy and the ot [illegible] by

In 1925 Calvé described a flattening of vertebral bodies [illegible]
sidered similar to Legge-Calvé-Perthe's disease of the femoral heads a
other osteochondritides. Buchman (1927) and Mezzari (1938), and later Massaro (1945) agreed that the etiology probably was an aseptic necrosis of the vertebral body resulting from diminution of the blood supply. Pathologic investigations disclosed fragmentation and collapse of the affected vertebral body. A propensity for regeneration of bone and gradual restoration of the vertebra to approximately its normal height was noted.

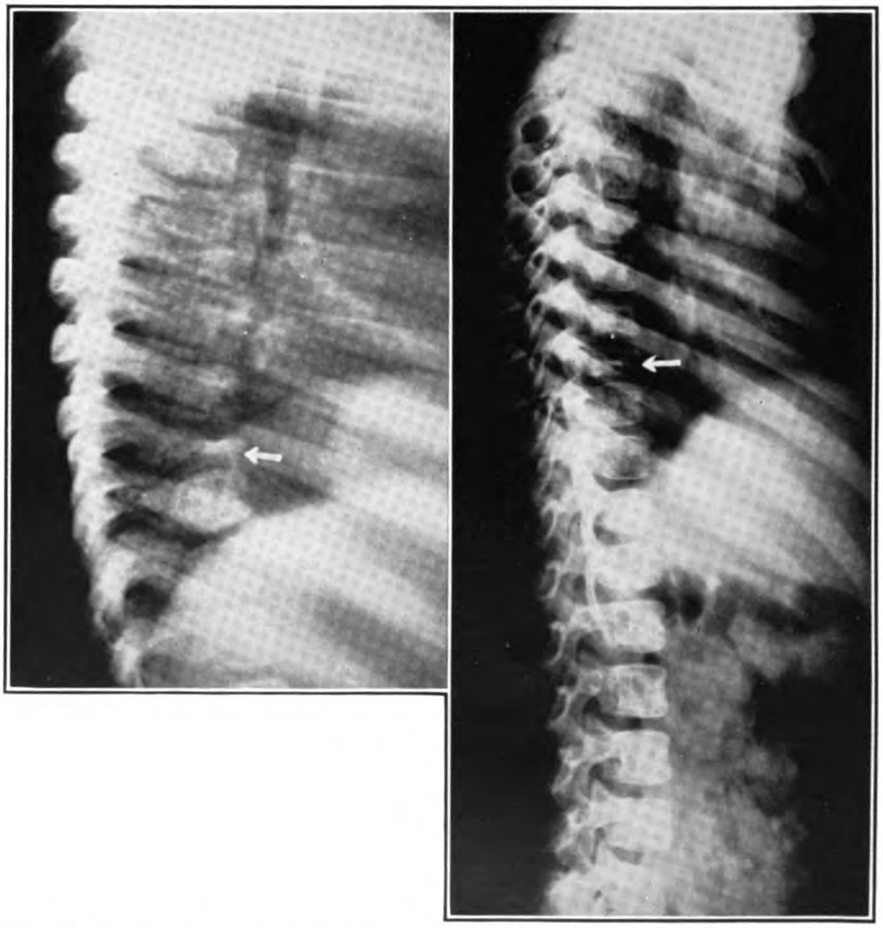

FIG. 385.—Nonlipoid reticulosis in a twenty-nine-month-old female. Collapse of the body of the ninth thoracic vertebra (arrow). This was followed by regrowth after treatment with folic acid antagonists, with marked relief of symptoms. (Courtesy of Dr. Robert S. Sherman and Dr. Sheldon Fox, Memorial Center, N. Y. C.)

In 1954 Compere, Johnson and Coventry advanced the concept that Calvé's disease actually was due to eosinophilic granuloma of the vertebral body, substantiating their impression with evidence obtained from vertebral body biopsy. This concept has gained general recognition, and the diagnosis of Calvé's disease is losing acceptance because of the frequency with which [illegible] the [illegible] characteristics of eosinophilic granuloma. However, [illegible] concept [illegible] osteochondritis of the vertebral body should not be dismissed entirely. The case reported by Rosselet (1949) would appear to qualify as an example [illegible] Calvé's disease.

Vertebra plana due to eosinophilic granuloma or possibly osteochondritis occurs mainly in children from about three to thirteen years of age. The associated symptoms may be only a mild backache and malaise, with some tenderness and spasm. The lesion may be symptomless, and identified only incidentally. A good response to *x*-ray therapy has been observed. Fripp (1958) reported that healing occurred spontaneously in four cases seen by him.

As a rule vertebra plana in children can be differentiated from other conditions which produce a similar malformation. Included in this group are conditions as diverse as lymphoma, Gaucher's disease, osteogenesis imperfecta, and other osteoporotic disturbances. The similarities and dissimilarities to spinal tuberculosis was discussed by Leeser (1947).

LETTERER-SIWE'S DISEASE

This is another of the nonlipoid granulomatous diseases of the reticulo-endothelial system. It occurs in infants, who rarely survive long enough to develop skeletal changes. The syndrome of Letterer-Siwe's disease is characterized by a diffuse, generalized hyperplasia of the reticulo-endothelial system with splenomegaly, hepatomegaly, a hemorrhagic tendency, generalized lymphadenopathy, generalized tumors overlying the bones, secondary anemia and increase in nonlipoid containing macrophages. The disease is of a relatively short duration with a fatal outcome (Siwe, 1933; Abt and Denenholtz, 1936; Dennis and Rosahn, 1951). Microscopically, the eosinophiles and macrophages are numerous, but foam cells are absent or relatively scant. Reports of vertebral extension of this disease are infrequent. Wallace (1949) encountered a collapsed first lumbar vertebra in a four-year-old boy with Letterer-Siwe's disease.

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GARGOYLISM (Dysostosis multiplex, liponchondrodystrophy, Hunter-Hurler disease, Pfaundler-Hurler syndrome.)

This is a rare familial disorder characterized by multiple symmetrical skeletal deformities with a superimposed disturbance of lipoid metabolism. In a report of the pathologic changes and biochemical alterations associated with gargoylism, Henderson, MacGregor, Thannhauser and Holden (1952) reported that almost all of the tissues of the body are affected by the same cellular lesion, in which individual cells showed cytoplasmic swelling with clear vacuolated or granular cytoplasm. An abnormal storage of a still unknown material, believed to be of a lipoid nature by some, and by others glycogen. These authors did not support either of these hypotheses, but suggested that a glycoprotein or mucoprotein might be the offending agent. Prior to this Hubeny and Delano (1941) suggested that this glycoprotein might be similar to the lipoid dystrophies such as Niemann-Pick's disease. Hervis (1950) mentioned that the parenchymal cells of the brain and liver were much more involved with a lipid, the exact nature of which was still under investigation. He, too, associated the disease with Niemann-Pick disease, Tay-Sachs disease and Gaucher's disease, in which the type of lipid was already known, as did Magee (1950).

Straus, Merliss and Reiser (1947) reviewed the literature and reported on a sixth case studied at necropsy. Chemical studies performed on this patient pointed towards a disease of collagenous connective tissue, chiefly a[illegible] the [illegible]d ligaments, rather than an abnormality of bony growth. [illegible] ept [illegible] was not a constant feature of the disease entity in their opinion [illegible] entirely [illegible]en present was found in the brain and in the reticulo-endothelial system. Chemical analysis of the tissues in their case presented a significant increase in the lipid content of the lymph nodes but not of the brain, spleen or liver. The increased lipid was by exclusion simple fat probably in a complex protein combination. The fact that it was not a phospholipid, cerebroside, or cholesterol separated this disturbance from the other idopathic lipoid dystrophies. In a later report Smith, Hempelmann, Moore and Barr (1952) reported that biochemical and pathologic studies indicated that the basic defect was a dysmetabolism of glycogen or an associated macromolecular substance. Histologic investigation showed increased glycogen-like substances in the liver and to a lesser extent in the pituitary, adrenal medulla and the bone marrow. Uzman (1955) concluded that gargoylism represented a genetically induced defect in the metabolism of structural polysaccharides.

Gargoylism usually is apparent at birth, but symptoms appear during the first or second years of life. In a review of 138 examples from the literature, to which 10 cases were added from his own experience, Jervis (1950) mentioned that 1 patient was normal until four years of age. This, however, is quite uncommon. In its complete form, a patient with gargoylism is mentally retarded, with a large head and short neck, an ugly face characterized by a flat nasal bridge, thick protuberant lips, delayed dentition and widely spaced and poorly developed teeth. The skull is deformed, its appearance simulating hypertelorism, scaphocephaly and macrocephaly. A characteristic bump appears in the immediate vicinity of the location of the anterior fontanelle. The eyes are set low and the eyebrows are coarse. While the infant may grow for a year or so, dwarfism and a lumbar kyphosis soon become evident. The mobility of the various articulations is reduced. The patient presents a prominent abdomen and short extremities with broad, short hands which may have a claw-like deformity. Accompanying this are other changes such as poor muscular development, hypertrichosis, leathery skin, deafness and a tendency towards umbilical hernia (Zellweger, Ciaccui and Firzli, 1952). Hepatomegaly and splenomegaly are frequently present. The cornea becomes cloudy due to replacement of Bowman's membrane by swollen cells distended with granular material and lipid laden swollen cells in the substantia propria (Lindsay, Reilly, Gotham and Skaher, 1948). These authors concluded from 8 necropsies, and biopsies from 4 other patients, that the granular intracellular material was composed largely of glycogen and glycoprotein.

Changes occur in the long bones, including swelling of the shafts of the humeri, tapering towards their ends, together with a tilting of the distal epiphyseal lines of the radius and ulna towards each other. Retardation of bone age development with broadened metacarpal and phalangeal bones are prominent. The ribs present a practically diagnostic appearance characterized by thinning and constriction of their proximal portions close to their spinal articulations, and gradual flaring distal to this site, particularly in the lower ribs. Thickening of the sternal ends of the clavicles and shallow and small glenoid and acetabular fossæ have been mentioned (Lahey, Lowman and Worth, 1947).

Interestingly enough, the bones of the lower extremities are less affected. Coxa vara hip deformities may appear. Caffey (1952) reported on ricket-like changes in the long bones in cases of gargoylism in the early stage of the disease, which disappeared spontaneously. He rega[illegible] her hi [illegible] to unrecognized sign of gargoylism, occurring during the p[illegible] liosis [illegible]ch the lipids or glycogen became deposited in both cortical and s[illegible] bone. This osteoporotic phase may have been overlooked pre[illegible] 38-*B*, because *x*-ray examinations were not available in the very early stages. [illegible] cases observed over a period from fifteen to twenty-four months, thoracolumbar kyphosis did not appear.

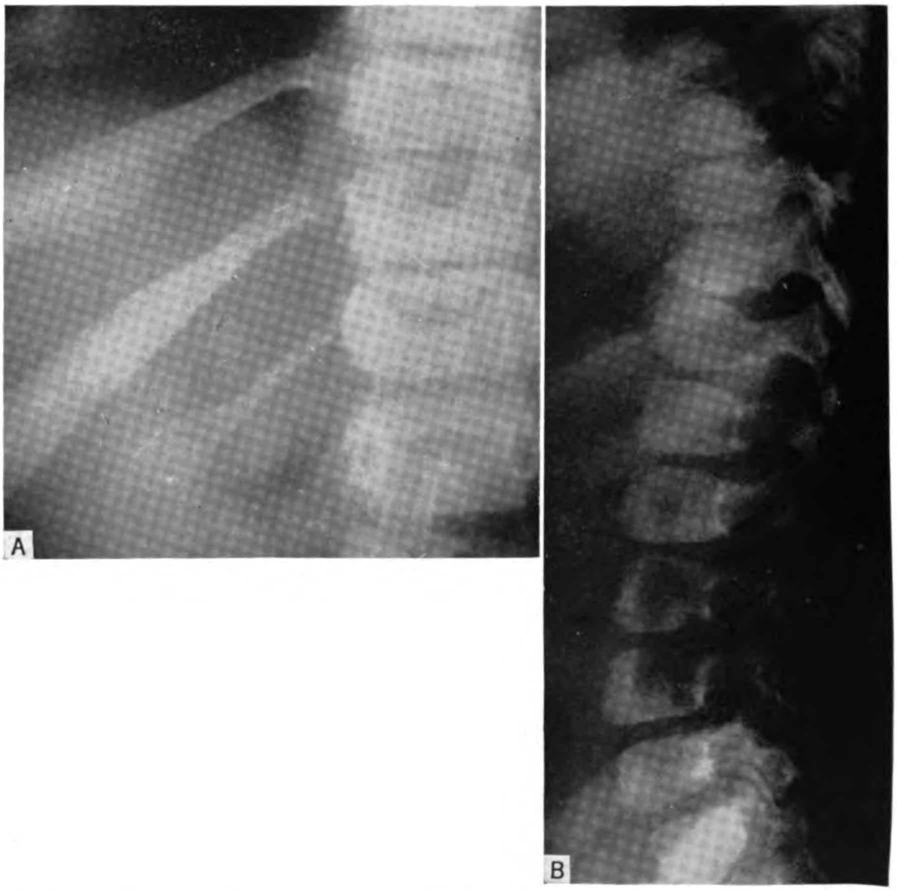

FIG. 386.—Gargoylism in a seventeen-month-old male infant. The proximal ends of the ribs present narrow, ribbon-like appearances. The vertebral bodies (*B*) show a tongue-like protrusion at the anterior aspect of the eleventh thoracic vertebra. The remaining vertebræ are rather small, have rounded edges and are somewhat asymmetric.

Involvement of the spine is considered one of the more frequent evidences of gargoylism. Kyphoscoliosis is observed quite frequently and is one of the more constant changes (Ellis, Sheldon and Capon, 1936). Fairbank, (1949) mentioned that the upper and lower surfaces of the vertebral bodies may present a circular rather than a quadrilateral appearance. In the presence of an angular kyphosis one body is usually smaller and more wedge-like than the others, and is displaced dorsally as if squeezed out of of line. Its anterior part is deficient and the lower portion projects forward in a beak-like manner. This usually occurs in the second lumbar vertebra, [illegible] firs[illegible] ve t[illegible] being next in frequency. The vertebral bodies [illegible]ove a[illegible]th retain a more or less normal appearance (Harvey, 1942; Larson a[illegible]ichty, 1943). Lurie and Levy (1944) attributed the vertebral anomalie[illegible] faulty formation of cartilage, and suggested that the retarded bone growth may be due to a pituitary factor. They noted that enlargement of the head and the dorsolumbar kyphosis appeared at approximately the same time between the first and second years of life, at which time growth seemed to stop and normal mental and physical development ceased. The involvement of the thoracolumbar spine was also commented on by Jervis (1950) who mentioned that the other vertebræ may also show irregularities of outline and variations in size. Thomson and Vesterdal (1951) attributed the deformity of the vertebræ and the resultant kyphosis to the absence of some of the limbus centers of ossification of the vertebral bodies.

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Chapter

10

Aortic Aneurysms

ANEURYSMS of the aorta usually are arteriosclerotic or syphilitic in origin. Those in the thoracic aorta are rather likely to be syphilitic, while abdominal aortic aneurysms are predominantly of the arteriosclerotic variety (Epstein, 1945). Arteriosclerotic aneurysms have a tendency to be of the diffuse fusiform type, while syphilitic aneurysms are more often of the sacculated and multiple variety.

Other factors infrequently associated with aneurysms are trauma, contiguous extra-arterial disease with secondary injury to the vascular walls, inflammatory vascular lesions and rheumatic fever. An unusual form of aneurysmal dilatation of the abdominal aorta with communication with

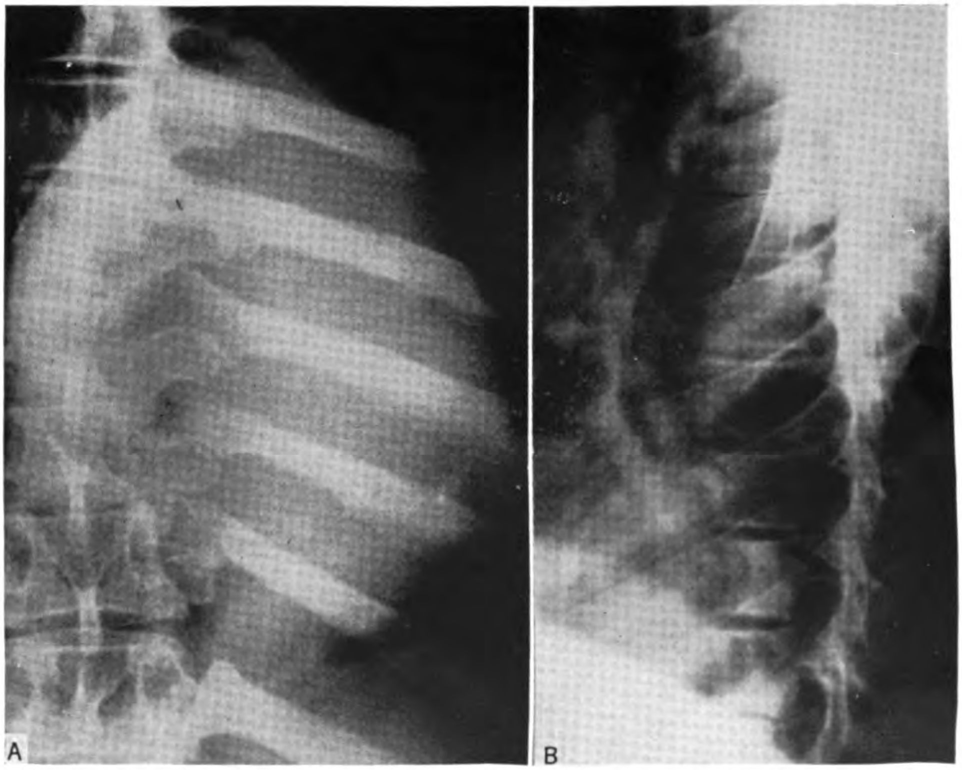

FIG. 387.—Aneurysm of descending thoracic aorta eroding the left lateral aspect of the mid-thoracic vertebræ and adjacent rib heads (*A*). Erosion of the anterior aspects of the lower thoracic vertebræ (*B*) in another case. Note the scalloped edges of the eroded vertebræ. The intervertebral discs are well maintained.

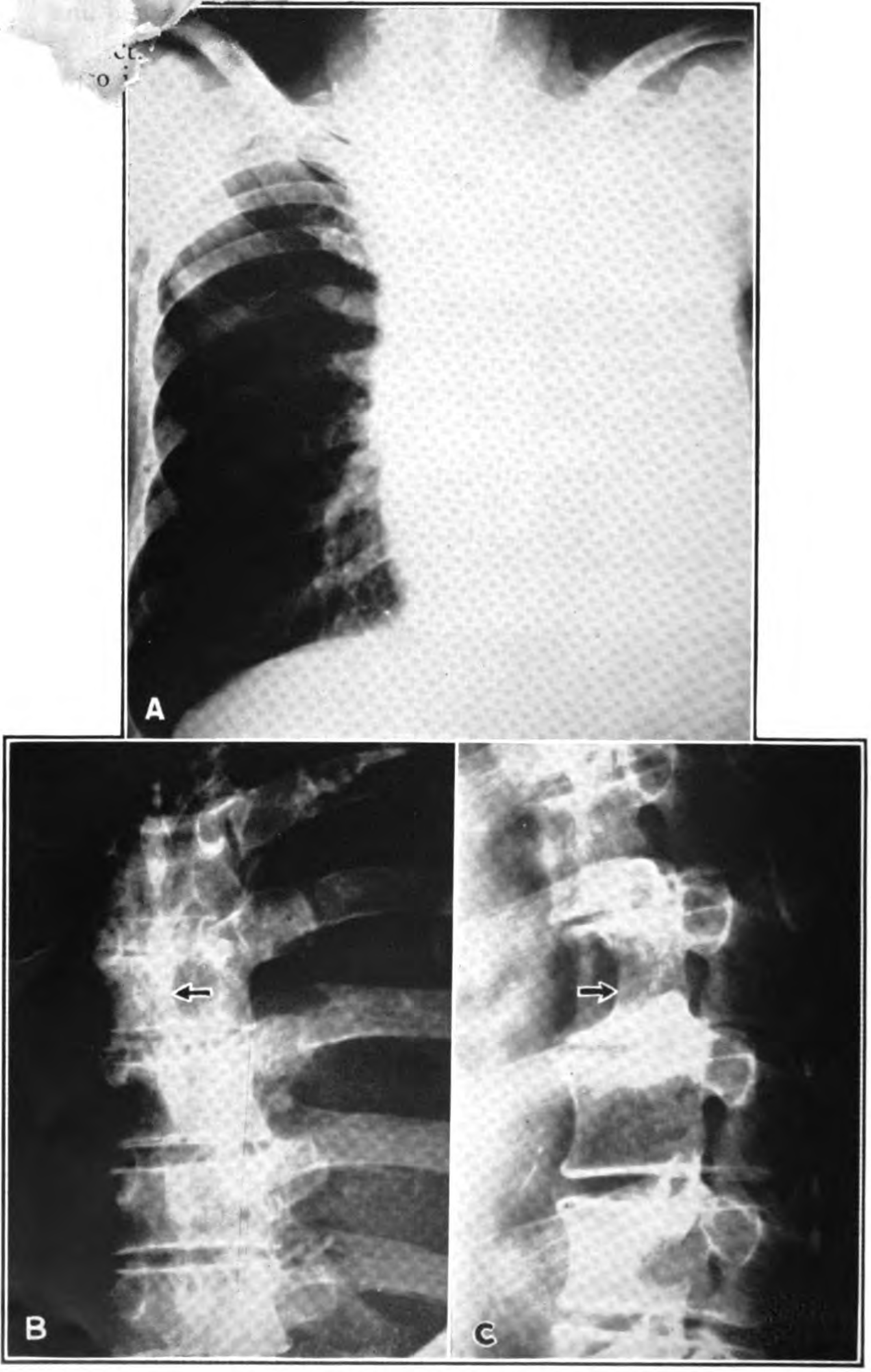

FIG. 388.—*A*, Aneurysm of the descending thoracic aorta obscured by overlying left pleural effusion. *B*, Same case, anteroposterior view of thoracic spine. A scalloped erosion is present on the left lateral aspect of the sixth thoracic vertebra (arrow). *C*, Same case, oblique view of thoracic spine. The scalloped area is indicated by the black arrow.

the abdominal venous circulation was describe [illegible] Azulay (1952). This occurred in a thirty-four-year- [illegible] a Corrigan pulse without clinical signs of aortic insufficie [illegible] rs were heard in the lower vertebral and posterior pelvic reg [illegible] Radiographic examination disclosed numerous lytic lesions in the [illegible] lumbar vertebra and mixed lytic and blastic lesions in the left sacrum and ilium. The radiologic diagnosis was angiosarcoma resulting in arteriovenous fistulas. Necropsy revealed a large aneurysmal sac occupying most of the posterior pelvic cavity. Extending posteriorly were dilated vessels which entered the pelvic and vertebral bones to form loculated interconnected intraosseous vascular channels which were considered developmental anomalies. This patient died in cardiac failure.

Aneurysms of the ascending and transverse thoracic aorta are not accompanied by vertebral changes because of their anatomic positions. Aneurysms of the descending thoracic aorta may produce erosive changes in the anterolateral aspects of the vertebræ against which they impinge. This constant compressive and pulsatile trauma produces smooth, scalloped erosions which are deepest in the midportions of the involved vertebræ. Such changes are more common in the lower thoracic vertebræ, but may occur at any level associated with aortic vertebral compression. They are usually multiple. The intervertebral discs, being avascular and relatively resilient, are not affected. Similar changes are produced in the upper lumbar vertebral bodies by abdominal aortic aneurysms. These are most frequent in the upper three lumbar vertebræ.

The changes may be identified roentgenologically on anteroposterior, oblique and lateral films. In the early stage the faint scalloping may be better seen on oblique films. The diagnosis may be facilitated by the identification of a soft tissue paravertebral mass, within which a linear calcification concave towards the spine may appear (Klinefelter, 1946). In some cases the aneurysm may be associated with obliteration of the psoas shadow and deflection of the spine contralaterally suggesting a perinephric abscess (Uhle, 1941). Abdominal aneurysms may intrude into the stomach or duodenum, simulating a neoplasm. Death may follow rupture of the aneurysm into the upper gastrointestinal tract. A case of aneurysm perforating into the third portion of the duodenum and eroding adjacent vertebræ had been reported as early as 1859 (Rottino, 1943). A large aneurysm of the descending thoracic aorta with retropsoas extension was reported by Putts and Bacon (1936).

Aneurysms of the renal artery are uncommon, and as a rule are not in relationship to the spine. They may sometimes be identified because of an opaque, ring-shaped shadow with a relatively dense periphery in the kidney pelvis region (Howard, Suby and Harberson, 1941). The diagnosis is best established by means of aortography.

Dissecting aneurysms of the aorta occur once in every 400 to 500 cases examined at necropsy (Moersch and Sayre, 1950). They occur more often in males, the ratio being from 2 to 3 males to each female. Those in the fifth to sixth decades of life appear to be more vulnerable, although no adult age group is entirely exempt. The usual clinical picture is one of sudden pain and rapid death. Survival for weeks or months may take place while the aneurysm dissects its way down from the thoracic region. In some the intimal tear may be at the proximal portion of the thoracic aorta, and death occur from exsanguination from a rupture of one of the iliac arteries.

[illegible] sms may be associated with spinal cord ischemia [illegible] sensory disturbances. Tuohy, Boman and Berde [illegible] inal cord ischemia results from interruption of blood [illegible]m the aorta. Moersch and Sayre ascribed the neurologic [illegible] to impairment of the blood supply to the cord because of interference with the intercostal arteries depriving the anterior spinal artery of its circulation. Scott and Sancetta (1949) gathered 28 cases of paraplegia and paraparesis resulting from dissecting aneurysm from the literature. They reported a fifty-six-year-old male in whom the intercostal arteries had been severed, causing hemorrhagic infarction of the cord and subarachnoid hemorrhage. The plain film roentgenograms and myelography were normal. In 6 cases of dissecting aneurysm seen by us, the *x*-ray examination of the spine was normal in all. None had neurologic manifestations, death having occurred quickly after the onset of the dissection.

With the advent of improved technics for the surgical correction of aortic aneurysms, neurologic complications after aortic surgery have been reported. Adams and van Geertruyden (1956) reported 24 instances of ischaemia of the spinal cord associated with aortic surgery from the literature and their own experience. They found that cord lesions were localized to the lower spinal cord, and the variations of clinical manifestations resulted from differences in the extent and degree of ischemia. Translumbar aortography also has been associated with paraplegia, attributed to the injection of neurotoxic contrast materials which reach the spinal cord in sufficient concentration to cause neurocellular damage.

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